ORIGINAL ARTICLE



# Prevalence Estimate of Cerebral Palsy in Northern Alberta: Births, 2008-2010

*Charlene M.T. Robertson, M. Florencia Ricci, Kathleen O'Grady, Maryam Oskoui, Helly Goez, Jerome Y. Yager, John C. Andersen*

**ABSTRACT:** ***Objectives:*** The objectives of this study were to determine prevalence estimates of cerebral palsy (CP) among 5-year-old children in northern Alberta; to provide congenital, gestational age– and birth weight–specific, and postneonatal CP rates; and to describe motor subtypes and function. ***Methods:*** This population-based prevalence estimate study, part of the Canadian Cerebral Palsy Registry, reports confirmed CP diagnoses at age 5 years made by pediatric rehabilitation and child neurology specialists. Prevalence rates with 95% confidence intervals (CIs) used Alberta government denominators of same-age children and live births. ***Results:*** The Northern Alberta CP rate (birth years, 2008-2010) for 173 5-year-old children is 2.22 (95% CI 2.12, 2.32) per 1000 5-year-old children. The congenital CP rate is 1.99 (95% CI, 1.89-2.09) per 1000 live births; unilateral congenital CP, 1.0 (95% CI, 0.64-1.36) per 1000 live births; and postneonatal CP, 0.12 (95% CI, 0.1-0.14) per 1000 live births. Gestational age-specific rates are similar: age <28 weeks, 27.2 (95% CI, 23.05-31.35) and 28 to 31 weeks, 29.5 (95% CI, 25.78-33.22). Motor subtypes for 169 children (data missing, 4; male, 97; postnatal, 9) are: spastic, 148 (87.6%) including 31 (20.9%) with diplegia, 10 (6.8%) triplegia, 33 (22.2%) quadriplegia, 74 (50%) hemiplegia/monoplegia); and dyskinetic, 18 (10.6%) and ataxic, 3 (1.8%). A total of 107 (63.3%) ambulate without assistive devices and 111(65.7%) handle most objects with their hands independently. ***Conclusions:*** This is the fourth Canadian CP prevalence study; one from Quebec used a similar case ascertainment approach and two 1980s studies from Alberta and British Columbia used administrative databases. Northern Alberta CP rates are comparable with other developed countries. The hemiplegic subtype is the most common. Rates among preterm children have declined but are similar for the <28 and 28 to 31 gestation-week groups.

**RÉSUMÉ: Estimations de la prévalence de la paralysie cérébrale chez les enfants du nord de l'Alberta nés entre 2008 et 2010.** ***Objectifs:*** Cette étude s'est donné trois objectifs : estimer la prévalence de la paralysie cérébrale (PC) chez les enfants âgés de 5 ans du nord de l'Alberta ; faire connaître les taux de PC de nature congénitale et post-néonatale en plus de fournir des données au sujet de l'âge gestationnel; et décrire les divers troubles de la mobilité qui y sont associés ainsi que leurs caractéristiques. ***Méthodes:*** Cette étude visant à estimer la prévalence de la PC s'est appuyée sur le Registre canadien de paralysie cérébrale. Elle repose donc sur des diagnostics établis par des spécialistes en réadaptation pédiatrique et en neurologie infantile chez des enfants de 5 ans. Les taux de prévalence (IC 95%) reposent sur des données du gouvernement albertain ayant permis de déterminer le nombre de sujets du même âge et de naissances vivantes. ***Résultats:*** Le taux de PC chez les enfants du nord de l'Alberta nés entre 2008 et 2010, ce qui représente 173 sujets âgés de 5 ans, est de 2,22 (IC 95 % ; 2,12 - 2,32) par tranche de 1000 enfants âgés de 5 ans. Le taux de PC congénitale était de 1,99 (IC 95 % ; 1,89 - 2,09) par tranche de 1000 naissances vivantes. Le taux de PC congénitale unilatérale était de 1,0 (IC 95 % ; 0,64 -1,36) par tranche de 1000 naissances vivantes. Enfin, il était de 0,12 (IC 95 % ; 0,1 – 0,14) par tranche de 1000 naissances vivantes en ce qui concerne la PC post-néonatale. Soulignons que les taux de PC ne varient guère en fonction de l'âge gestationnel : âge < 28 semaines : 27,2 (IC 95 % ; 23,05 – 31,35) ; 28 à 31 semaines : 29,5 (IC 95 % ; 25,78 – 33,22). Sur un total de 169 enfants, les données étant incomplètes pour les 4 autres, on a pu identifier les troubles de la mobilité suivants: spastiques (148, soit 87,6 %), ce qui incluait 31 sujets atteints de diplégie (20,9 %), 10 de triplégie (6,8 %), 33 de quadriplégie (22,2 %) et 74 d'hémiplégie/monoplégie (50 %); dyskinétiques (18, soit 10,6 %) ; et ataxiques (3, soit 1,8 %). Fait à noter, 97 sujets étaient des garçons ; 9 sujets ont aussi vu leur trouble apparaître à la suite de leur naissance. Un total de 107 sujets (63,3 %) pouvaient se déplacer sans recourir à des appareils et à des accessoires fonctionnels tandis que 111 (65,7 %) étaient en mesure de manipuler de façon autonome des objets courants avec leurs mains. ***Conclusions:*** Notre étude constitue la quatrième étude canadienne portant sur la prévalence de la PC. Une étude menée au Québec avait utilisé la même approche de détermination de cas. Deux autres études, conduites dans les années 80 en Alberta et en Colombie-Britannique, avaient plutôt utilisé des bases de données administratives. Chose certaine, les taux de prévalence de la PC dans le nord de l'Alberta sont comparables à ceux des autres pays développés. Le trouble de la mobilité le plus répandu s'est avéré l'hémiplégie. Enfin, les taux de PC chez les enfants prématurés ont décliné ; ils demeurent néanmoins analogues dans le cas d'enfants dont l'âge gestationnel était inférieur à 28 semaines ou variait entre 28 et 31 semaines.

**Keywords:** cerebral palsy, children, prevalence estimate, population-based, hemiplegia

doi:10.1017/cjn.2017.33 Can J Neurol Sci. 2017; 44: 366-374

From the Department of Pediatrics, University of Alberta, Edmonton, Alberta, Canada (CMTR, MFR, HG, JYY, JCA); Pediatric Rehabilitation, Glenrose Rehabilitation Hospital, Edmonton, Alberta, Canada (KO); Department of Pediatrics and Neurology & Neurosurgery, McGill University, Montreal, Quebec, Canada (MO)

Received September 1, 2016. Final Revisions Submitted December 20, 2016. Date of Acceptance January 13, 2017.

*Correspondence to*: Charlene M. T. Robertson, Glenrose Rehabilitation Hospital, 10230-111th Ave, Edmonton, Alberta, Canada, T5G 0B7. Email: Charlene.robertson@ahs.ca

Cerebral palsy (CP) is the leading cause of childhood-onset, lifelong physical disability in the Western world.[1] CP is "a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication, and behaviour; by epilepsy, and by secondary musculoskeletal problems."[2,3] Motor sub-types of CP include spastic (bilateral, unilateral (hemiplegia, monoplegia)), dyskinetic (dystonic, choreoathetotic), ataxic, and mixed.[3-5] Twenty-four months is accepted as the maximum age that an acquired injury to the developing brain may be considered as CP.[6]

A systematic review and meta-analysis of the median prevalence estimate of CP for birth years 1985 through 2004, with studies published by 2006, gives a pooled rate of 2.11 per 1000 live births (95% confidence interval [CI], 1.98-2.25). [7] Rates in Western countries rose from the birth years of the 1960s to peak in the 1990s and dropped in the 2000s,[8-12] with a decrease in diplegia and an increase in hemiplegia over this time[8,9]; risk factors included low-birth-weight survival and multiple birth.[8] The increase in the mid-1990s prevalence of CP associated with an increase in survival of extremely preterm infants was followed by a stabilization and decline in this at-risk group.[10-13] It is not known if the overall prevalence in northern Alberta has changed since the 1985 through 1988 birth years when the rate from administrative databases was 2.57 (95% CI, 2.41-2.79) per 1000 living children at age 8 years.[14]

In general, there are two approaches determining the prevalence of CP, administrative databases and population-based registries (e.g. data from patient registries), with no significant differences in rates as yet determined between the two methods.[15] The Canadian Cerebral Palsy Registry is a population-based registry, expanded from the *Registre de la paralysie cerebrale du Quebec*, as part of the NeuroDevNet National Networks of Centres of Excellence CP Demonstration Project.[7,15] The first report from The Canadian CP Registry shows an overall rate per 1000 children alive at 9-10 years of 1.84 (95% CI, 1.6-2.08) using the frequentist approach. This study from northern Alberta gives the second report from the Canadian CP Cerebral Palsy Registry.

The primary study objective is to determine the overall population-based prevalence estimate of CP among 5-year-old children in northern Alberta for the birth years 2008 through 2010. Secondary objectives are to provide congenital and postneonatal CP rates and frequency among multiple births and by gestational age– and birth weight–specific subgroups and to describe the motor subtypes and function of these children at age 5 years.

## Methods

### The Study Region

Northern Alberta comprises about two-thirds of the geographic region of the Province of Alberta (661,185 $km^2$) and for 2008 through 2010 had 49% of 151,405 live births.[16,17] Of live births, infant deaths occurred in 0.62%, preterm births in 8.7%, and multiple births in 3.5%. Alberta has universal-access government-funded health care with two regional perinatal/neonatal programs (north and south) for at-risk mothers and ill newborns. In northern Alberta, one centrally located rehabilitation center, the Glenrose Rehabilitation Hospital, a tertiary care center with four associated outreach clinics, assesses and treats almost all children with CP.[14]

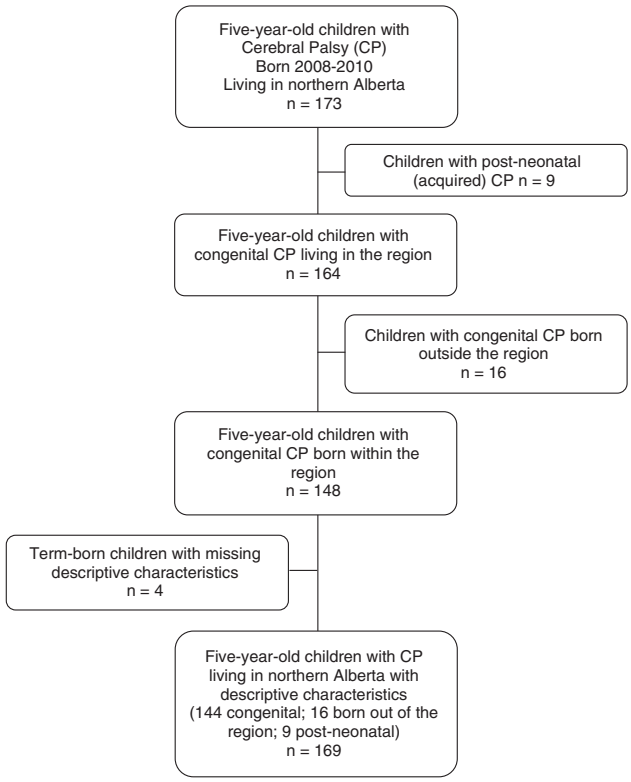


***Figure 1:*** *Flow sheet describing 173 children with cerebral palsy, born 2008-2010.*

### Subjects

The study population includes 173 children (Figure 1), with a confirmed diagnosis of CP at age 5 years who reside in northern Alberta, were born between January 2008 and December 2010, and are registered for care at the Glenrose Rehabilitation Hospital. Children with suspect CP are referred from the community, other hospitals, and the Neonatal Follow-up Clinic. CP diagnoses are made by specialists in pediatric rehabilitation and child neurology. Motor subtype classifications follow standard practice.[3-5] Exclusion criteria include any child whose home is not northern Alberta even if they receive Glenrose rehabilitation services and any child without a diagnosis of CP at age 5 years. Although children with CP from southern Alberta are now part of the Canadian CP Registry, they were added later and are not part of this study.[6]

### Registration, Case Ascertainment, and Data Extraction

As part of the Canadian CP Registry, participants are ascertained after their second birthday and diagnosis confirmed after their fifth birthday. At the time of initial registration, data were collected from maternal medical and obstetrical records and the child's neonatal, medical, and rehabilitation charts. Data collection forms were initially created for the Quebec CP Registry through a Delphi consensus process using a panel of provincial, national, and international experts to determine definitions, motor subtype



https://doi.org/10.1017/cjn.2017.33 Published online by Cambridge University Press

classifications, and maternal and child-related variables to be collected.[6] Subjects' health records were reviewed again at 5 years of age for confirmatory evidence of the diagnosis of CP and functional classification. If confirmation of diagnosis was not available, the subject was evaluated by a child neurologist or pediatric rehabilitation specialist and the confirmed CP diagnosis recorded.

### Ethics

This study was approved by the Health Research Ethics Board, Health Panel, University of Alberta, Edmonton, Alberta, with waiver of parental/guardian consent.

## Measures

### Population-Based Prevalence Estimate Study

#### *Case Ascertainment*

Through a long-standing referral system, diagnosis and care for children with CP in northern Alberta is regionalized. Case ascertainment is considered to be high, based in part, upon our previous documentation in the administration database study[14] showing all but 1.1% of children with CP in this region were registered at the Glenrose Rehabilitation Hospital.

#### *Prevalence*

For the years 2008 to 2010, overall and yearly prevalence estimates of CP for 5-year-old children per 1000 children of all same age living in the same region regardless of place of birth were established as well as the rates per 1000 live births of congenital and postneonatal CP, frequency per among multiples, and gestational age– and birth weight–specific groups.

#### *Denominator*

A live birth is defined as "a complete expulsion or extraction from the mother, irrespective of the duration of the pregnancy, of a fetus in which, after expulsion or extraction, there is breathing, beating of the heart, pulsation of the umbilical cord or definite movement of a voluntary muscle."[17] The number of live births in northern Alberta from 2008 to 2010 was 74,173, corresponding to regional referral patterns from Capital, East Central, Aspen, Peace Country, Northern Lights, and one-third of the David Thomson regions.[16,17] Born between 2008 and 2010, there were 77,779 5-year-old children living between 2013 and 2015 in northern Alberta, exceeding live births because of immigration numbers.

#### *Numerators*

The overall numerator for the birth years 2008 through 2010 is the sum of all the individually identified 5-year-old children living in northern Alberta with confirmed CP, whether born in or outside of northern Alberta. This was further defined by timing of possible cause of CP as congenital or postneonatal and by gestational-age and birth-weight groups.

#### *Descriptive Variables*

Variables include hospital of birth; year of birth; estimated time of occurrence of CP, fetal, and ≤28 days of life (congenital) or postneonatal (with the cause of the final diagnosis of CP happening after 28 days and before 2 years of life); sex; maternal age; type of pregnancy, single or multiple birth; type of delivery, vaginal or caesarean section; birth weight (grams); and gestational age (completed weeks).

CP motor subtype and topography[4,5] as well as motor function classification is reported. The definitions of the motor subtypes of CP used in this study include: (1) spastic diplegia, predominantly involving the two lower extremities where there may be some upper extremity involvement but any upper limb tone abnormality is less than in the lower limbs; (2) spastic triplegia, predominately involving three extremities, including both lower extremities and one upper extremity, or leg-dominant spasticity, or both upper extremities and one lower extremity, or arm-dominant spasticity; (3) spastic quadriplegia, involving all four extremities and including double hemiplegia in which the involvement of the arms is equal or more than of the legs; (4) spastic hemiplegia, which refers to only one hemibody part, including involvement of only one limb (monoplegia); (5) dyskinetic, which includes any combination of dystonia or involuntary (choreoathetotic) movement; and (6) ataxia, which includes loss of muscular coordination where movements have abnormal force, rhythm, and accuracy.[4,5] Functional mobility is described according to The Gross Motor Function Classification System (GMFCS)[18] and typical performance of hand use is described according to the Manual Ability Classification System (MACS).[19]

### Statistical Analysis

Continuous variables are presented as means (standard deviation) and categorical variables as counts and percentages. The prevalence estimates of CP based on frequentist approach[15] is given per 1000 children of the same age and per cohort-specific live birth using 95% CIs. Deidentified data were collected and managed using REDCap[20] electronic data capture tools hosted and supported by the Women and Children's Health Research Institute at the University of Alberta and transferred to SPSS, version 21, for presentation and analysis.

## Results

### Prevalence Estimates of CP

For the birth years 2008 through 2010, there were 173 5-year-old children living in northern Alberta with confirmed CP diagnoses giving an overall rate for the 3-year period of 2.22 (95% CI, 2.12-2.32) per 1000 living children age 5 years. Table 1 gives the characteristics of 169 of the 173 children, but does not include four term-born children (one born in 2008, three in 2010) with confirmed CP but missing descriptive characteristics. Of the 169, 57% are boys, 15% were one of a multiple birth, 39% were <2500 grams at birth, and 38% were born prematurely. Two other children born in the same period and receiving rehabilitation services at Glenrose Rehabilitation Hospital are not included in these rates. One child had spastic right CP at 2 years of age, but the diagnosis was not confirmed at 5 years; another was diagnosed with a degenerative neurological disease. There were no known deaths from the time of registration until reporting at 5 years.

Yearly rates and cohort-specific rates are found in Table 2. Excluding nine (5.2%) children considered to have postneonatal CP causation, the prevalence for congenital CP based on 164 children at age 5 years with detailed data is 2.1 (95% CI, 2.0-2.2) per 1000 children living in northern Alberta at age 5 years. The rate for congenital CP per 1000 live births is 1.99 (95% CI,



https://doi.org/10.1017/cjn.2017.33 Published online by Cambridge University Press

**Table 1: Characteristics of 5-year-old children with CP born in 2008-2010 and living in northern Alberta in relation to location of birth**

| Characteristics | Total (n = 169)* | Location of Birth | | |
|---|---|---|---|---|
| | | Northern Alberta (n = 153)* | | Outside of northern Alberta (n = 16) |
| | | Congenital (n = 144)* | Post-neonatal (n = 9) | |
| Date of birth (year), n (%) | | | | |
| 2008 | 60 (35.5) | 51 (35.4) | 4 (44.4) | 5 (31.2) |
| 2009 | 59 (34.9) | 51 (35.4) | 1 (11.1) | 7 (43.8) |
| 2010 | 50 (29.6) | 42 (29.2) | 4 (44.4) | 4 (25) |
| Male, n (%) | 97 (57.4) | 81 (56.3) | 5 (55.6) | 11 (69) |
| Maternal age (years), n (%) | | | | |
| 15-18 | 11 (6.5) | 11 (7.6) | 0 (0) | 0 (0) |
| 19-25 | 49 (29) | 40 (27.8) | 3 (33.3) | 6 (37.5) |
| 25-35 | 94 (55.6) | 82 (56.4) | 6 (66.7) | 6 (37.5) |
| 35-40 | 12 (7.1) | 10 (6.9) | 0 (0) | 2 (12.5) |
| > 40 | 1 (0.6) | 1 (0.7%) | 0 (0) | 0 (0) |
| Unknown | 2 (1.2) | 0 (0) | 0 (0) | 2 (12.5) |
| Type of pregnancy, n (%) | | | | |
| Single fetus | 143 (84.6) | 121 (84) | 8 (84.6) | 14 (87.5) |
| Type of delivery, n (%) | | | | |
| Vaginal | 85 (50.3) | 69 (47.9) | 5 (55.6) | 11 (68.8) |
| Caesarean section | 81 (47.9) | 73 (50.7) | 4 (44.4) | 4 (25) |
| Unknown | 3 (1.8) | 2 (1.4) | 0 (0) | 1 (6.3) |
| Birth weight (grams), n (%) | | | | |
| < 1000 | 13 (7.7) | 12 (8.3) | 0 (0) | 1 (6.3) |
| 1000-1499 | 16 (9.5) | 14 (9.7) | 1 (11.1) | 1 (6.3) |
| 1500-2499 | 34 (21.1) | 28 (19.4) | 2 (22.2) | 4 (25) |
| ≥ 2500 | 103 (60.8) | 90 (62.5) | 6 (66.7) | 7 (43.8) |
| Unknown | 3 (1.8) | 0 (0) | 0 (0) | 3 (18.8) |
| Gestational age (weeks), n (%) | | | | |
| < 28 | 13 (7.7) | 12 (8.3) | 1 (11.1) | 0 (0) |
| 28-31 | 22 (13) | 17 (11.8) | 0 (0) | 5 (31.3) |
| 32-36 | 30 (17.8) | 29 (19) | 2 (22.2%) | 1 (6.3) |
| ≥ 37 | 103 (60.9) | 88 (61.1) | 6 (66.7) | 9 (56.3) |
| Unknown | 1 (0.6) | 0 (0%) | 0 (0) | 1 (6.3) |

*Excludes four children born at term with confirmed CP with missing descriptive data.

1.89-2.09). For those born as a twin or triplet, the multiple birth–specific rate is 8.99 (95% CI, 7.88-10.1) per 1000 multiple live births. The rate for unilateral congenital CP is 1.0 (95% CI, 0.64-1.36) per 1000 live births. Table 2 also summarizes the CP rates for Alberta for birth years 1985 through 1988 from an administrative database.[14]

### Postneonatal Timing of Possible Causes of CP

Nine children had postneonatal causes for CP (Tables 1 and 2). Causes determined by neurology consultation included: accidental head injury, 2; nonaccidental head injury (shaken baby syndrome), 2; congenital complications of congenital heart defect/cerebrovascular accident secondary to cardiac arrest, 2; cerebrovascular accident (perisurgical), 1; postconvulsion anoxia, 1; and infection (meningitis), 1. Of these nine, five had direct neonatal risk factors for CP.

### Motor Type and Topography

Table 3 gives the motor subtypes for 169 children with available characteristics in relation to gestational age and birth-weight subgroups. There were no differences in the proportion of CP subtypes in relation to birth within or outside of northern Alberta.



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**Table 2: CP in northern Alberta: birth years, 2008-2010, and in Alberta, birth years 1985-1988: presentation of prevalence estimate by different study approaches**

| | Birth years | |
|---|---|---|
| | **2008-2010** | **1985-1988*** |
| Geographic region | Northern Alberta | Alberta |
| Study approach | Case ascertainment | Administrative database |
| Age of children (years) | 5 | 8 |
| Number of children with CP | 173 | 248 |
| Population of same-age children | 77,779 | 96,359 |
| Prevalence estimate per same-age children | 2.22 (95% CI, 2.12-2.32) | 2.57 (95% CI, 2.41-2.79) |
| Year 1 | 2.38 (95% CI, 2.19-2.57) | 2.71 (95% CI, 2.13-3.26) |
| Year 2 | 2.26 (95% CI, 2.08-2.44) | 2.28 (95% CI, 1.77-2.79) |
| Year 3 | 1.89 (95% CI, 1.73-2.05) | 2.72 (95% CI, 2.45-2.99) |
| Frequency per 1000 live births by birth weight groups, grams | | |
| < 1000 | 40.1 (95% CI, 34.5-45.7) | 98.4 (95% CI, 59.5-137.4) |
| 1000-1499 | 22.7 (95% CI, 19.3-26.0) | 70.7 (95% CI, 43.8-97.6) |
| 1500-2499 | 6.7 (95% CI, 5.9-7.5) | 11.0 (95% CI, 7.9-14.1) |
| ≥ 2500 | 1.3 (95% CI, 1.2-1.4) | 1.7 (95% CI, 1.6-1.8) |
| Frequency per 1000 live births by gestational age groups, weeks | | |
| < 28 | 27.2 (95% CI, 23.05-31.35) | NA |
| 28-31 | 29.5 (95% CI, 25.78-33.22) | NA |
| 32-36 | 5.01 (95% CI, 4.43-5.59) | NA |
| ≥ 37 | 1.29 (95% CI, 1.21-1.37) | NA |
| Number of children with postnatal CP | 9 | 19 |
| Prevalence estimate of post-natal CP per 1000 live births | 0.12 (95% CI, 0.1-0.14) | 0.2 (95% CI, 0.17-0.23) |

*Data from reference 14.
NA = not available.

Motor subtypes includes spasticity, 148 (87.6%) of which 31 (20.9%) had diplegia, 10 (6.8%) triplegia, 33 (22.2%) quadriplegia, 74 (50%) hemiplegia/monoplegia; 18 were dyskinetic (10.6%); and three were ataxic (1.8%). Term births were recorded for 10 (32.3%) of those with spastic diplegic CP, 25 (58.1%) with spastic tri- or quadriplegia, 52 (70.2%) with hemiplegia, and 14 (77.7%) with dyskinetic CP. Of the nine with postneonatal CP, three had spastic quadriplegia, two were dyskinetic, and four had hemiplegic CP. Unilateral CP comprises 43.8% of the 169 with documented characteristics, 46.3% of all congenital CP, and 51.3% of congenital CP born in this region.

## Function

Functional mobility relative to CP type is shown on Table 4. Of 169 children with confirmed CP, 107 (63.3%) can walk without need for a hand-held mobility device indoors and outdoors (n = 79) or for short distances on level surfaces outdoors (n = 28) (GMFCS level I/II); 17 (10.1%) use a hand-held mobility device and need assistance for ambulation for uneven terrain and stairs (GMFCS III), whereas in 45 (26.6%) self-mobility is limited, predominately using a manual wheelchair or powered mobility (GMFCS IV/V). Of those 105 with spastic diplegic or hemiplegic CP, 95 (90.5%) have mobility within level I/II.

Hand function is given in Table 4. Of 169 children with confirmed CP, 111 (65.7%) could perform activities with their hands without the need for modification or preparation, although alternative ways of performance might be needed (MACS I/II); 20 (11.8%) handled objects with difficulty and needed activities to be adapted to their needs (MACS III); and 38 (22.5%) required continuous support or total assistance (MACS IV/V). Of the 105 with spastic diplegic or hemiplegic CP, 99 (94.2%) function within MACS level I/II.

Of the nine children with postneonatal CP, five function at level V for both the GMFCS and MACS.

## Discussion

Because of differences in measurements of the numerator and denominator, the age at diagnosis, the inclusion of those with postneonatal causes, statistical approaches, and possible death before diagnosis, comparisons of the prevalence of CP remain difficult.[5,21] For northern Alberta, we have shown an overall population-based prevalence estimate of CP, based on confirmed diagnoses after the children reached 5 years of age, with the denominator per 1000 same-age children from the same period to be within the usually quoted prevalence rate of 2 to 2.5 per 1000 live births. Our rate is similar to that reported in the recently determined pooled rate[7] and by the Australian Cerebral Palsy



https://doi.org/10.1017/cjn.2017.33 Published online by Cambridge University Press

**Table 3: Gestational age groups in relation to motor type and topography for 169 5-year-old children with confirmed CP born 2008-2010***

| CP type | Number (%) of total | Gestational age groups and numbers | |
|---|---|---|---|
| | | Gestational age Groups (weeks) | n (%) within groups for each type |
| Spastic CP: 2 extremities predominantly involved (diplegia) | 31 (18.3) | <28 | 1 (3) |
| | | 28-31 | 12 (39) |
| | | 32-36 | 8 (26) |
| | | ≥37 | 10 (32) |
| Spastic CP: 3 extremities predominantly involved (triplegia) | 10 (5.9) | <28 | 1 (10) |
| | | 28-31 | 3 (30) |
| | | 32-36 | 0 (0) |
| | | ≥37 | 6 (60) |
| Spastic CP: 4 extremities predominantly involved (quadriplegia) | 33 (19.5) | <28 | 4 (12) |
| | | 28-31 | 2 (6) |
| | | 32-36 | 8 (24) |
| | | ≥37 | 19 (58) |
| Ataxic CP | 3 (1.8) | <28 | 1 (33) |
| | | 28-31 | 0 (0) |
| | | 32-36 | 0 (0) |
| | | ≥37 | 2 (67) |
| Dyskinetic CP: predominantly dystonic | 8 (4.7) | <28 | 1 (12.5) |
| | | 28-31 | 0 (0) |
| | | 32-36 | 1 (12.5) |
| | | ≥37 | 6 (75) |
| Dyskinetic CP: predominantly choreoathetotic | 10 (5.9) | <28 | 0 (0) |
| | | 28-31 | 1 (10) |
| | | 32-36 | 1 (10) |
| | | ≥37 | 8 (80) |
| Spastic CP: right hemiplegia | 36 (21.3) | <28 | 3 (8) |
| | | 28-31 | 1 (3) |
| | | 32-36 | 4 (11) |
| | | ≥37 | 28 (78) |
| Spastic CP: left hemiplegia/monoplegia† | 38 (22.6) | <28 | 2 (5) |
| | | 28-31 | 3 (8) |
| | | 32-36 | 8 (21) |
| | | ≥37 | 24 (63) |
| | | Unknown | 1 (3) |

*Excludes 4 children born at term with confirmed CP with missing descriptive data.
†1 child had monoplegia.

Register.[11] Our study has a number of similarities with the Australian report, including rate of postneonatally determined CP and proportion of boys, preterm births, and plurality. [11]

Although comparison of rates remains problematic, Oskoui et al,[7] in a large systematic review, has shown no significant difference in overall prevalence between population-based data and administrative data.[7] Our presentation of the current results in Table 2, beside our administrative database results of more than 20 years ago, show no overlap in the 95% CIs for the studies. This suggests improvement at least in the rates of CP among the low-birth-weight children; for these children, we are confident in complete case ascertainment because of the complete follow-up by the Neonatal Follow-up Program in northern Alberta.[13] This study also suggests a trend toward lower rates than the British Columbia study using administrative data.[27] Our prior linkage study used administrative diagnostic codes that may not be comparable; on the other hand, the denominator for both studies included children of the same age as those with diagnosed CP, rather than live births. The prevalence of this present study may be an underestimate. The final year of this study shows a lower rate,



https://doi.org/10.1017/cjn.2017.33 Published online by Cambridge University Press

**Table 4: Functional mobility of 169 5-year-old children with confirmed CP in relation to motor type and topography: birth years 2008-2010***

| CP type | Number (%) of total | Level of functional mobility | Functional mobility n (%) | |
|---|---|---|---|---|
| | | | GMFCS | MACS |
| Spastic CP: 2 extremities predominantly involved (diplegia) | 31 (18.3) | I | 17 (54.8) | 27 (87.1) |
| | | II | 8 (25.8) | 3 (9.7) |
| | | III | 5 (16.1) | 1 (3.7) |
| | | IV | 1 (3.2) | 0 (0) |
| | | V | 0 (0) | 0 (0) |
| Spastic CP: 3 extremities predominantly involved (triplegia) | 10 (5.9) | I | 1 (10) | 2 (20) |
| | | II | 2 (20) | 2 (20) |
| | | III | 4 (40) | 4 (40) |
| | | IV | 2 (20) | 2 (20) |
| | | V | 1 (10) | 0 (0) |
| Spastic CP: 4 extremities predominantly involved (quadriplegia) | 33 (19.5) | I | 0 (0) | 0 (0) |
| | | II | 1 (3) | 1 (3) |
| | | III | 6 (18.2) | 10 (33.3) |
| | | IV | 8 (24.2) | 3 (9.1) |
| | | V | 18 (54.5) | 19 (57.6) |
| Ataxic CP | 3 (1.8) | I | 1 (33.3) | 2 (66.7) |
| | | II | 0 (0) | 0 (0) |
| | | III | 1 (33.3) | 0 (0) |
| | | IV | 0 (0) | 0 (0) |
| | | V | 1 (33.3) | 1 (33.3) |
| Dyskinetic CP predominantly dystonic | 8 (4.7) | I | 2 (25) | 4 (50) |
| | | II | 3 (37.5) | 1 (12.5) |
| | | III | 0 (0) | 0 (0) |
| | | IV | 0 (0) | 0 (0) |
| | | V | 3 (37.5) | 3 (37.5) |
| Dyskinetic CP, predominantly choreoathetotic | 10 (5.9) | I | 0 (0) | 0 (0) |
| | | II | 2 (20) | 0 (0) |
| | | III | 0 (0) | 2 (20) |
| | | IV | 1 (10) | 2 (20) |
| | | V | 7 (70) | 6 (60) |
| Spastic CP: right hemiplegia | 36 (21.3) | I | 0 (83.3) | 20 (55.6) |
| | | II | 4 (11.1) | 13 (36.1) |
| | | III | 1 (2.8) | 2 (5.6) |
| | | IV | 0 (0) | 0 (0) |
| | | V | 1 (2.8) | 1 (2.8) |
| Spastic CP: left hemiplegia/monoplegia† | 38 (22.6) | I | 28 (73.7) | 22 (57.9) |
| | | II | 8 (21.1) | 14 (36.8) |
| | | III | 0 (0) | 1 (2.6) |
| | | IV | 2 (5.3) | 1 (2.6) |
| | | V | 0 (0) | 0 (0) |
| All CP | | I | 79 (46.7) | 77 (45.6) |
| | | II | 28 (16.6) | 34 (20.1) |
| | | III | 17 (10.1) | 20 (11.8) |
| | | IV | 14 (8.3) | 8 (4.7) |
| | | V | 31 (18.3) | 30 (17.8) |

*Excludes 4 children born at term with confirmed CP with missing descriptive data.
†1 child had monoplegia.



https://doi.org/10.1017/cjn.2017.33 Published online by Cambridge University Press

1.89 (95% CI, 1.73-2.05), suggesting possible incomplete ascertainment relative to the first 2 years. We have no explanation for this apparent improvement other than year-to-year variability.

Our study supports the current reported change in the idea of an inverse relationship between gestational age and CP rates with proportionately fewer extremely low-birth-weight children diagnosed with CP.[4,8,10-12,22] Similar to others,[7,10] we have shown a rate of CP within the extremely preterm children that is lower than those of 28 to 31 weeks' gestation. This reduction is unlikely the result of chance.[12] The birth weight–specific rates presented here are lower than those we gave for births in 1985 through 1988,[14] but higher than we reported for children of 20 to 27 weeks' gestation born between 2001 and 2003.[13] Variability in the recording of live births for the lowest gestational age and birth weight subgroups[15] presents difficulty for the comparison of gestation- and birth weight–specific prevalence rates.

Postneonatal CP rates are lower in developed countries, decreasing over the past three decades.[21,23] We report these rates as lower than our previous report,[14] similar to the mean European rate, but higher than other current rates.[10,23]

Heterogeneity of reported motor CP subtypes has resulted in calls to make this classification more reliable.[24] Compared with the Victorian study from birth years 1970 through 2003,[24] our study shows the proportion of children with dyskinesia to be more than double, with lower rates of ataxia and slightly lower rates of spasticity. For some time, there have been concerns about an increasing rate of unilateral CP.[8,9] Although this increase has not been linear,[12] the proportion of unilateral spastic CP in relation to all subtypes is increasing.[12] We are reporting one of the highest rates of unilateral spastic CP, which makes up half of all spastic CP in this study. Similar findings of other investigators are leading to increased investigation of perinatal stroke.[25]

Neurological CP subtyping has been found to be a strong predictor of functional status related to ambulation.[26] Compared with the Quebec study,[26] fewer children in our study have GMFCS levels of IV/V. Differences in GMFCS levels for similar subtypes may be explained in part by other comorbidities not addressed by this report.[26] We have shown that 65.7% of all children and 94.3% of those with spastic diplegia and hemiplegia/monoplegia have MACS levels of I/II.

A strength of this study is case ascertainment, with a confirmed diagnosis for children older than age 5 years giving high true positives. However, because complete case ascertainment is critical to the numerator, this is also a limitation of the study as only those referred to the Glenrose Rehabilitation Hospital and its satellites are included. Previous documentation reported that all but 1.1% of children with CP for this region were assessed at Glenrose[13]; however, our results may still underestimate the true rates in this geographically large area. Although this study uses the methodology for one of the two approaches to the study of CP prevalence estimates recently outlined,[15] case ascertainment would be stronger if multiple sources had been used rather than information from a single tertiary center and associated satellites. This region's population is smaller with fewer cases relative to larger registries,[10-12] reducing the generalizability of results, particularly for subgroups. A strength is the denominator of the number of children of the same age as the children with CP, allowing for the most appropriate denominator for the overall prevalence estimate. A limitation is the denominator of live births; using the current definition, the number of live births is larger than the previous definition that included only live births with gestational ages ≥20 weeks and birth weights ≥500 grams. The present denominator is larger and hence the prevalence rates are slightly lower than if previous cutoff levels were used. Similarly, denominators based upon neonatal survivors, if available, would give somewhat higher prevalence rates and assist with comparison with other studies. Limitations of this study also include having no information on deceased children with likely CP before age 2 years of age and omission of children leaving the area. We have not addressed comorbidities. Comparison with current administrative data from the same years would be an asset.

## Conclusion

The population-based prevalence estimate of CP in northern Alberta is within the current levels for the Western world and suggests a reduction in congenital CP prevalence relative to our previous study from the 1980s birth years, especially for the low-birth-weight children. Rates for unilateral spastic CP make up the most common subtype. The gestational age–specific rate for those with CP of <28 weeks is similar to that of those of 28 through 31 weeks. The results provide needed regional information.

## Acknowledgements and Funding

We thank the Canadian Cerebral Palsy Registry for funding, direction, and outline for data collection that has made this study possible. We thank the referring doctors of Northern Alberta for their continued early referral of young children with suspect cerebral palsy to pediatric rehabilitation at the Glenrose Rehabilitation Hospital and for the devoted disciplines at Glenrose that care for these children and make these data available for study.

The Canadian Cerebral Palsy Registry has been funded by the NeuroDevNet Networks of Centres of Excellence, Ottawa, Ontario, Canada, and the generous support of the Stollery Children's Hospital Foundation through the Women and Children's Health Research Institute and the Glenrose Rehabilitation Hospital Foundation, Edmonton, Alberta, Canada. These funding agencies have not been part of the design, data collection, data analysis, manuscript preparation, or publications decisions for this manuscript.

## Disclosures

MO has the following disclosures: funding recipient, Kids Brain Health Network (Networks of Centres of Excellence), Ottawa, ON, Canada; consultant and consulting fees: Biogen. JCA has the following disclosures: principal investigator, research grant, Kids Brain Health Network (NeuroDevNet) NCE; Women's and Children Health Research Institute; funding recipient, Glenrose Rehabilitation Hospital Foundation, Edmonton, Alberta, Canada. CMTR, MFR, KO, HG, and JYY do not have anything to disclose.

## Statement of Authorship

CMTR completed a literature search; wrote and revised the manuscript; participated in the concept, design, writing the main document, revising, and approval of the final version; and had access to the original database. MFR prepared a report for one of the funding agencies, the Women and Children's Health Research Institute, University of Alberta, Edmonton, Alberta, Canada, outlining the progress of the study; she also submitted an abstract

to the NeuroDevNet fall meeting; and had access to the data and participated in the interpretation of analysis and the revision and final versions of the manuscript. KO collected and entered the data to REDCap and hence had access to all data and was part of the original design of the study, revising and approving of the final version. MO represents the Canadian Cerebral Palsy Registry on this manuscript and she revised and approved the final version. HG was part of the original study design, revising and approval of the final manuscript. JYY was part of the original study design, reading and approving the manuscript. JCC completed all aspects of the study including concept/design, supervision of collection of outcomes data, interpretation of analysis, and revisions of the original and final versions of the manuscript; he represents the Canadian Cerebral Palsy Registry on this article.

Following is the list of site primary investigators of the Canadian Cerebral Palsy Registry.

Michael I. Shevell, MD, and Maryam Oskoui, MD, MSc (Department of Neurology/Neurosurgery, McGill University; Division of Pediatric Neurology, McGill University Health Centre; Department of Pediatrics, McGill University); John Andersen, MD (Developmental Pediatrics, University of Alberta; Glenrose Rehabilitation Hospital); David Buckley, MD (Department of Pediatrics, Memorial University); Darcy Fehlings, MD, MSc (Institute of Health Policy, Management and Evaluation, University of Toronto; Dalla Lana School of Public Health, University of Toronto; Department of Pediatrics, University of Toronto; Division of Developmental Pediatrics, Holland Bloorview Kids Rehabilitation Hospital and Bloorview Research Institute); Adam Kirton, MD, MSc (Departments of Pediatrics and Clinical Neurosciences, University of Calgary; Alberta Children's Hospital Research Institute); Alison J. Moore, MD, MSc (Department of Pediatrics, University of Calgary; Developmental Pediatrics and Neurology Alberta Children's Hospital); Nicole Pigeon, MD (Department of Pediatrics, Centre hospitalier universitaire de Sherbrooke; Centre de réadaptation Estrie); Louise Koclas, MD (Division of Neonatology, McGill University Health Centre; Centre de Réadaptation Marie Enfant, CHU Sainte Justine); Esias van Rensburg, MD (Department of Pediatrics, University of British Columbia; Neuromotor Program and Division of Developmental Pediatrics, Sunny Hill Health Centre for Children); Ellen Wood, MD MSc (Faculty of Medicine, Dalhousie University; Department of Pediatrics, IWK Health Centre).

## References


1. Hirtz D, Thurman DJ, Gwinn-Hardy K, Mohamed M, Chaudhuri AR, Zalutsky R. How common are the 'common' neurological disorders? Neurology. 2007;68:326-37.
2. Rosenbaum P, Paneth N, Leviton A, Goldstein M, Bax M. A report: the definition and classification of cerebral palsy. Dev Med Child Neurol. 2007;49(supp109):8-14.
3. Smithers-Sheedy H, Badawi N, Blair E, et al. What constitutes cerebral palsy in the twenty-first century. Dev Med Child Neurol. 2014;56:323-8.
4. SCPE Collaborative Group. Surveillance of cerebral palsy in Europe; a collaboration of cerebral palsy surveys and registers. Dev Med Child Neurol. 2000;42:816-24.
5. McIntyre S, Morgan C, Walker K, Novak I. Cerebral palsy—don't delay. Development Disabil Res Re. 2011;17:114-29.
6. Shevell M, Dagenais L, Oskoui M. The epidemiology of cerebral palsy: new perspectives from A Canadian Registry. Semin Pediatr Neurol. 2013;20:60-4.
7. Oskoui M, Coutinho F, Dykaman J, Jette N, Pringsheim T. An update on the prevalence of cerebral palsy: a systemic review and meta-analysis. Dev Med Child Neurol. 2013;55:509-19.
8. Odding E, Roebroeck ME, Stam HJ. The epidemiology of cerebral palsy: incidence, impairments and risk factors. Disabil Rehabil. 2006;28:183-91.
9. Ravn SH, Flachs EM, Uldall P. Cerebral palsy in eastern Denmark: declining birth prevalence but increasing numbers of unilateral cerebral palsy in birth year period 1986-1998. Eur J Paediatr Neurol. 2010;14:214-8.
10. Reid SM, Meehan E, McIntyre S, Goldsmith S, Badawi N, Reddihough DS. on behalf of the Australian Cerebral Palsy Register Group. Temporal trends in cerebral palsy by impairment severity and birth gestation. Dev Med Child Neurol. 2016; 58(Supp 2):25-35.
11. Smithers-Sheedy H, McIntyre S, Gibson C, et al. on behalf of the Australian Cerebral Palsy Register Group. A special supplement: findings from the Australian cerebral palsy Register, birth years 1993 to 2006. Dev Med Child Neurol. 2016;58(Supp 2):5-10.
12. Sellier E, Platt MJ, Andersen GL, Krageloh-Mann I, De La Cruz J, Cans C. on behalf of Surveillance of Cerebral Palsy Network. Decreasing prevalence in cerebral palsy: a multi-site European population-based study, 1980 to 2003. Dev Med Child Neurol. 2016;58:85-92.
13. Robertson CMT, Watt M-J, Yasui Y. Changes in the prevalence of cerebral palsy in children born very prematurely within a population-based program over 30 years. JAMA. 2007;297:2733-40.
14. Robertson CMT, Svenson LW, Joffres MR. Prevalence of cerebral palsy in Alberta. Can J Neurol Sci. 1998;25:117-22.
15. Oskoui M, Joseph L, Dagenais L, Shevell M. Prevalence of cerebral palsy in Quebec: alternative approaches. Neuroepidemiology. 2013;40:264-8.
16. Government of Alberta. Interactive Health Data Application. Available at: http://www.ahw.gov.ab.ca/IHDA.
17. Male and female live births, birth weight by gestational period, 2008-2010. Edmonton, Alberta, Canada: Government Services, Alberta Vital Statistics, Annual Reviews, 2008-2010; Tables 4, 7, and 8.
18. Palisano R, Rosenbaum P, Walter S, Russell D, Wood E, Galuppi B. Development and reliability of a system to classify gross motor function in children with cerebral palsy. Dev med Child Neurol. 1997;39:214-23.
19. Eliasson A-C, Krumlinde-Sundholm L, Rosblad B, et al. The Manual Ability Classification System (MACS) for children with cerebral palsy: scale development and evidence of validity and reliability. Dev Med Child Neurol. 2006;48:549-54.
20. Harris PA, Taylor R, Thielke R, Payne J, Gonzalez N, Conde JG. Research electronic data capture (REDCap) – A metadata-driven methodology and workflow process for providing translational research informatics support. J Biomed Inform. 2009;42:377-81.
21. Blair E, Watson L. Epidemiology of cerebral palsy. Semin Fetal Neonatal Med. 2006;11:117-25.
22. Himpens E, Van den Broeck C, Oostra A, Calders P, Vanhaesebrouck P. Prevalence, type, distribution, and severity of cerebral palsy in relation to gestational age: a meta-analytic review. Dev Med Child Neurol. 2008;50:334-40.
23. Germany L, Ehlinger V, Klapouszczak D, et al. Trends in prevalence and characteristics of post-neonatal cerebral palsy cases: a European registry-based study. Res Development Disabi. 2013;34:1669-77.
24. Reid S, Carlin JB, Reddihough D. Distribution of motor types in cerebral palsy: how do registry data compare. Dev Med Child Neurol. 2011;53:233-8.
25. Golomb MR, Garg BP, Saha C, Azzouz F, Williams LS. Cerebral palsy after perinatal ischemic stroke. J Child Neurol. 2008;23:279-86.
26. Shevell MI, Dagenais l, Hall N. The relationship of cerebral palsy subtype and functional motor impairment: a population-based study. Dev Med Child Neurol. 2009;51:872-7.
27. Smith L, Kelly KD, Prkachin G, Voaklander DC. The prevalence of cerebral palsy in British Columbia, 1991-1995. Can J Neurol Sci. 2008;35:342-7.