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Prevalence of Cerebral Palsy in Quebec: Alternative Approaches

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Prevalence · Registry · Cerebral palsy · Canada

Abstract
Aim: To provide an estimate of the period prevalence of cerebral palsy (CP) in the province of Quebec. ***Methods:*** Children with CP were identified from three consecutive birth cohorts (1999–2001) from the Quebec CP Registry, covering 6 of the 17 administrative health regions of the province. Two inferential approaches were applied for period prevalence estimation, frequentist and bayesian. ***Results:*** 228 children were identified with CP. Using a frequentist approach, the overall prevalence of CP was 1.84 per 1,000 children aged 9–11 years living in those areas in 2010 (95% CI 1.60–2.08). Using a bayesian approach taking into account the uncertainty about the registry's sensitivity in capturing all cases, the overall prevalence is higher at 2.30 per 1,000 children with a 95% CI (1.99–2.65). ***Conclusion:*** Using a bayesian approach to adjust for the registry's known high specificity and lower sensitivity, the prevalence estimate is in concordance with worldwide estimates and estimates using administrative databases in western Canadian provinces. Future studies are needed to validate the diagnosis of CP within administrative databases and to evaluate possible regional trends across Canada in both prevalence and health service utilization, which may highlight disparities in healthcare delivery.

Cerebral palsy (CP) is the most common cause of physical impairment encountered in children with an estimated prevalence of 2.0–2.5 per 1,000 live births in Western populations [1]. Studies drawn from a variety of geographically dispersed population-based service registries, including a recent study in California, suggest a clear temporal trend towards increasing survival well into adulthood [2]. Longevity is inversely correlated with disability, with most individuals without severe disabilities living to age 35 years and beyond [3]. As a chronic disorder across the lifespan, with a multiplicity of functional limitations and co-morbidities, it is not surprising that CP has substantial additional societal costs related to additional medical, rehabilitation and educational needs attached to each individual case [4]. Anticipating health service needs should ideally be based on current local prevalence estimates.

The overall prevalence of CP in Canada has only been studied in the provinces of British Columbia (BC) and Alberta using administrative datasets. In BC, a record search of administrative databases for the birth cohorts 1991–1995 was conducted using the ICD-9 CP diagnostic code '343' for case ascertainment recorded after 3 years of age [5]. The reported period prevalence was 2.68 per 1,000 live births. In Alberta, administrative databases were searched using similar inclusion and exclusion criteria as were used in BC [6]. The ICD-9 code '343' was also used for case ascertainment either recorded at 3 years of age or prior to 3

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years if recorded at least three times. The period prevalence was 2.57 per 1,000 children alive at 8 years. Four Canadian studies have also examined the prevalence of CP among cohorts of preterm infants Alberta, Nova Scotia, BC and Quebec [7–10]. There are no overall prevalence figures for CP in Quebec. The goal of this study is to provide an estimate of the period prevalence of CP in Quebec.

Methods

Case Ascertainment

The Multiregional Canadian population-based CP Registry was first established in Quebec as the Registre de la Paralyse Cérébrale de Québec (REPACQ). It is now being expanded through a variety of funding support to other provinces in Canada including Alberta, Ontario, BC, Nova Scotia and Newfoundland. Utilizing the framework of the regionalization of pediatric rehabilitation service delivery, children with CP born between 1999 and 2001 were enrolled within 6 of the province's 17 administrative health regions capturing 52.4% of all children aged 9–11 years living in Quebec as of 2010. The six administrative regions include: Capitale-Nationale, Estrie, Montreal, Outaouais, Lanaudière and Laurentides. In the province of Quebec, pediatric neurologists, developmental pediatricians and physiatrists working in the hospital settings systematically refer children with CP to regional rehabilitation centers as soon as the diagnosis has been established. Thus, ascertainment for the CP Registry was mainly carried out in the rehabilitation centers. Children with CP were identified by physicians and clinical coordinators working at the CP clinics of the regional rehabilitation centers. The list of children compiled from these clinics was compared to the list of children seen at each center by all rehabilitation professionals for completeness, including physical therapists, occupational therapists, speech therapists, psychologists and audiologists. The ascertainment strategies and timeline of registration and follow-up for the registry are outlined in online supplementary figure 1 (see www.karger.com/doi/10.1159/000345120). Overall, the mean number of ascertainment sources per child was 3, the first being the CP clinics and the second most common source being the lists from different professionals working with the children.

Ethical permission for the Registry's establishment and implementation has been obtained at the local host institution (McGill University Health Center Research Institute) and each participating pediatric rehabilitation center. Once children were identified, parents or guardians were approached for consent to participate in the Registry. Non-participants are also indexed by gender and region of residence for prevalence estimations. Participants in REPACQ are ascertained only when a child is beyond the age of 2 years and confirmed after 5 years of age. This avoids the potential false 'capture' of young infants with transitory neuromotor findings under 2 years of age [11]. The diagnosis is confirmed after 5 years of age based on medial record review at each participating rehabilitation center, with direct communication by the on-site research coordinator with the treating team for any clarification needed. The research coordinators are often members of the treating team. CP is defined in accordance with recent consensus statements as a non-progressive motor impairment of early onset, that is presumably cerebral in origin, which may or may not be associated with developmental delays, cognitive disability, language impairment, epilepsy, sensory (auditory or visual) loss, orthopedic abnormalities, or behavioral difficulties [12].

Measures of Functional Severity

The Gross Motor Function Classification System (GMFCS) is a 5-level classification system that describes the gross motor function of children and youth with CP, with well-established validity and reliability [13]. Distinctions between levels are based on functional abilities and the need for assistive technology with levels I–III being ambulant and levels IV–V being non-ambulant. The GMFCS level for each child was recorded at 2 years of age and reassessed when possible at 5 years of age. The most recent score was used in our analysis. The Manual Ability Classification System (MACS) is a newer 5-level classification system that describes the typical fine motor function for children with CP, with established validity and reliability [14]. Distinctions between the levels are based on the child's ability to handle objects and their need for assistance or adaptations to perform manual tasks in everyday life, ranging from level I with no restriction on activities of daily living to level V where total assistant is needed for activities of daily living. The MACS became available during the follow-up period of the registry and scores are reported only when available at 5 years of age.

Statistical Analyses

For this study, cases of CP (both participants and non-participants) were identified from the REPACQ registry born between January 1, 1999 and December 31, 2001. A dataset was compiled consisting of the number of individuals with CP within each of the 6 administrative health regions.

PASW statistics software version 18.0 (PASW, Chicago, Ill., USA, 2009) was used for data entry and frequentist statistical analysis, and the statistical software WINBUGS version 1.4.3 was used for bayesian analysis [15]. Two inferential approaches were applied for period prevalence estimation, frequentist and bayesian. The frequentist approach estimates the prevalence of CP using the number of cases as the numerator and the number of children of the same age living in the area as the denominator, based on 2010 Canadian census data for each region. Prevalence within each administrative health region, as well as overall, was estimated with 95% confidence interval (CI). This approach, however, assumes that all identified cases are true positives and represent all the true positives within the population.

The bayesian approach more realistically does not assume that the registry provides perfect diagnostic information, leading to the possibility of both false positive and false negative ascertainment of CP status. This approach starts with prior probability distributions for all unknown parameters (such as prevalence, the sensitivity and specificity of the CP diagnosis entry in the registry) and uses the data to update these parameters through the likelihood function into a posterior probability. The posterior probability represents what a researcher should believe after seeing the current dataset, and accounting for any information input through the prior distributions [16–18]. A uniform prior distribution [$\beta(1,1)$], which says all possible values are equally likely a priori, was used for the prevalence of CP, so that the final inferences are based on the data and not on any prior knowledge about the prevalence. Prior knowledge about the sensitivity and specificity of the diagnosis of CP within the registry was taken into consideration

and prior distributions were determined by an expert panel consensus (two neurologists and one physical therapist leading the registry) well aware of the strengths and limitations of the registry. A prior distribution was set for a very high specificity of >99.99%, represented by a uniform prior density ranging from 99.99 to 99.999999%, and a sensitivity range of 75–85%, represented by a β (194.03754–47.79375) prior density. To evaluate robustness of our estimates, two additional sensitivity priors were used: a 'pessimistic' range from 65 to 75% represented by a β (224.65295–95.73538) prior density, and an 'optimistic' range from 85 to 95% represented by a β (116.06404–12.04506) prior density.

Results

A total of 228 children with CP were identified across all three birth cohorts, of which 186 fully participated in the registry. Follow-up information and reassessment after 5 years of age was available on all 186 of the participating children. Boys made up 53.9% of all children with CP. Information on GMFCS level was available on all 186 participating children. The majority had GMFCS level of I (48%), with 130 (70%) having an ambulant GMFCS level (I–III). The MACS were available only on 137 children in the registry, with a third having a level I and the remaining groups being equally divided. The characteristics of these children are shown in table 1.

Per the frequentist analysis, the overall prevalence of CP across all 6 regions for all three birth cohorts combined was 1.84 per 1,000 children living in those areas in 2010 (95% CI 1.60–2.08). The period prevalence estimates per administrative health region show important regional differences (table 2), with the lowest prevalence estimate in the Laurentians, Lanudiere and Outaouais regions and higher estimates in Estrie, Capitale-nationale, and Montreal regions. These differences could not be accounted for by differences in population sizes for children 9–11 years of age.

Using a bayesian approach taking into account the uncertainty about the registry's sensitivity in capturing all cases, the overall prevalence is higher at 2.30 per 1,000 children 9–11 years with a 95% credible interval (CrI) (1.99–2.65). This higher estimate reflects the imperfect sensitivity accounting for the presence of false negative cases. To evaluate the robustness of this estimate, the prevalence estimate using the 'pessimistic' range of sensitivity allowing for a greater number of false negatives resulted in a higher prevalence of 2.63 per 1,000 children aged 9–11 years with a 95% CrI (2.26–3.04). The prevalence estimate using the 'optimistic' sensitivity range allowing for a smaller number of false negatives resulted in a lower prevalence of 2.04 per 1,000 children aged 9–11 years with a 95% CrI (1.76–2.35).

Table 1. Characteristics of children in REPACQ

	1999	2000	2001	Total (% available)
CP at 5 years, n[a]	77	75	76	228
Refusals, n[b]	13	15	14	42
Boys, n (%)[c]	41 (53)	44 (59)	38 (50)	123 (54)
GMFCS, n				
I	38	21	30	89 (48)
II	7	7	5	19 (10)
III	7	5	10	22 (12)
IV	10	9	8	26 (14)
V	9	11	10	30 (16)
MACS, n				
I	19	10	15	44 (32)
II	5	10	9	24 (17)
III	11	4	5	20 (15)
IV	6	9	8	23 (17)
V	10	8	8	26 (19)

[a] Number of children identified with CP at 5 years of age (participants reassessed at 5 years and non-participants).
[b] Number of non-participants.
[c] Number of boys (percentage of total number of children identified with CP).

Discussion

This study provides the first estimates of the overall prevalence of CP in Quebec, and the first population-based estimates in Canada. The two previous overall CP prevalence estimates in Canada were reported using different denominators, making comparison between the two more challenging [5, 6]. Using a live birth denominator when the numerator is being captured during a later time period (after 2 years of age in CP) can be misleading if migration of the population and infant mortality are not accounted for. This effect is more pronounced among premature newborns that have a higher mortality rate. Furthermore, the two previous Canadian studies were based on administrative databases which are increasingly being used in epidemiological studies. Administrative data sources have the benefit of being readily available at minimal cost and allow access to longitudinal data. However, they are primarily established for health service reimbursement and are not designed as accurate sources of clinical information. Furthermore, the diagnosis of CP within administrative databases has never been validated. Prevalence estimates for CP using administrative databases have been reported using only a frequentist approach, with misleading narrow CIs for the point estimate when

Table 2. REPACQ regional prevalence of CP (children alive in 2010)

Health region	CP, n[a]	Children alive 9–11 years in 2010, n[b]	Frequentist approach		Bayesian approach	
			prevalence per 1,000	95% CI per 1,000	prevalence per 1,000	95% CrI per 1,000
Capitale-Nationale	29	17,549	1.65	1.05–2.25	2.13	1.42–2.97
Estrie	24	9,487	2.53	1.52–3.54	3.28	2.11–4.69
Montréal	124	52,154	2.38	1.96–2.80	2.99	2.45–3.58
Outaouais	15	11,722	1.30	0.65–1.95	1.70	0.97–2.63
Lanaudière	18	15,050	1.20	0.65–1.75	1.57	0.93–2.36
Laurentides	18	17,899	1.01	0.55–1.48	1.32	0.78–1.99
Total	228	123,861	1.84	1.60–2.08	2.30	1.99–2.65

Values are based on the Canadian 2010 census data.
[a] Number of children with CP identified in REPACQ. [b] Number of children aged 9–11 years living in the same regions as per the 2010 Canada census.

very large population denominators are used. Using a bayesian approach allows researchers to intuitively take into account some of the uncertainty about their data. This approach has been used in prevalence studies on a number of different conditions but has not been applied to CP [16].

The bayesian prevalence estimates from the registry fall within the 2.0–2.5 per 1,000 children reported uniformly worldwide and within the estimates from administrative databases in western Canadian provinces. Although all cases in the registry are expected to be true positives (high specificity), it is suspected that all cases of CP within these regions have not been captured (variable sensitivity). Using a bayesian approach to adjust for this, the estimated prevalence is higher reflecting the uncertainty in complete case ascertainment. Regional differences can reflect variations in sensitivity (incomplete capture in some regions), or reflect a tendency for families to relocate to larger urban areas with more service access for their child's profile of impairments. Over a third of children in the registry had a GMFCS level within the ambulant range, with an expected long-term survival well into adulthood. There were 42 non-participating children with CP in our study, and they were included in our numerator for overall prevalence estimation. The limitations brought by this include an overestimating of the prevalence if indeed some of these children were found to have a different diagnosis, or if these children moved out of these regions.

Better characterization of the potential misclassification within the REPACQ registry (possible missed cases – sensitivity) would help target improvements for case ascertainment. Future studies are also needed to validate the diagnosis of CP within existing administrative databases such as physician billing claims and to assess the feasibility of using such databases for case ascertainment by comparing these with a patient registry. The specificity and sensitivity of the diagnosis of CP within administrative databases has not been evaluated. It will also be important to evaluate possible regional trends across Canada in both prevalence and health service utilization, which may highlight disparities in healthcare delivery. The Canadian Multiregional CP Registry will offer this opportunity once enrollment is complete in all areas, with comparison possible with administrative databases available provincially through the Canadian system of universal health coverage.

Adjusting for misclassification is essential for obtaining accurate prevalence estimates, which is needed to plan future health service delivery to individuals and their families with CP. Only with accurate and reliable data can planning be rational and comprehensive for both current and future expectations and needs.

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Disclosure Statement

The authors have no conflicts of interest to disclose.

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