J. Paediatr. Child Health (2005) **41**, 479–483

Cerebral palsy in Victoria: Motor types, topography and gross motor function

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Objectives: To study the relationships between motor type, topographical distribution and gross motor function in a large, population-based cohort of children with cerebral palsy (CP), from the State of Victoria, and compare this cohort to similar cohorts from other countries.

Methods: An inception cohort was generated from the Victorian Cerebral Palsy Register (VCPR) for the birth years 1990–1992. Demographic information, motor types and topographical distribution were obtained from the register and supplemented by grading gross motor function according to the Gross Motor Function Classification System (GMFCS).

Results: Complete data were obtained on 323 (86%) of 374 children in the cohort. Gross motor function varied from GMFCS level I (35%) to GMFCS level V (18%) and was similar in distribution to a contemporaneous Swedish cohort. There was a fairly even distribution across the topographical distributions of hemiplegia (35%), diplegia (28%) and quadriplegia (37%) with a large majority of young people having the spastic motor type (86%).

Conclusions: The VCPR is ideal for population-based studies of gross motor function in children with CP. Gross motor function is similar in populations of children with CP in developed countries but the comparison of motor types and topographical distribution is difficult because of lack of consensus with classification systems. Use of the GMFCS provides a valid and reproducible method for clinicians to describe gross motor function in children with CP using a universal language.

Key words: cerebral palsy; classification system; gross motor function; motor skills; motor type; population register; topographical distribution.

Cerebral palsy (CP) is the most common cause of physical disability affecting children in Australia and most developed countries, with a prevalence of approximately 2 per 1000 live births.[1] It has been described as 'an umbrella term covering a group of non-progressive, but often changing, motor impairment syndromes secondary to lesions or anomalies of the brain arising in the early stages of its development'.[2] Although these clinical syndromes are often not pure, recognition of the dominant motor types and topography has been important for research into causal pathways and possible prevention, correlation with brain imaging, and for establishing a prognosis and setting management goals and strategies. It may also trigger a search for associated problems such as epilepsy, feeding difficulties and silent hip subluxation, which may significantly impact on the well-being and development of these children.[3,4] Traditionally, CP has been classified according to motor type, topographical distribution and functional severity,[1–3] but as yet there has been no consensus reached on either the descriptors or the definitions of motor type and topographical distribution.

The motor type is usually described as spastic, dyskinetic, ataxic, hypotonic or mixed. Currently in the USA, under the auspices of the National Institutes for Health, a taskforce on childhood motor disorders is working on the important issue of motor type classifications and some helpful guidelines have been published.[5–7] The Surveillance of CP in Europe has also gone through a consensus process to develop standard definitions and classifications of topography and motor type with the use of a training CD-ROM. However, although classical presentations are easily recognized, there are many children with mixed or changing motor types that are difficult to define.[1,8]

Classifications according to topographical distribution are widely employed. Although hemiplegia, diplegia and quadriplegia are commonly used terms, monoplegia and triplegia sometimes exist as separate entities or may be grouped with hemiplegia and quadriplegia, respectively.[1] The expressions double hemiplegia, spastic tetraplegia and total body involvement are sometimes used to describe a child with four-limb involvement and with the upper limbs more severely involved than the lower limbs. Not surprisingly, classifications based on motor type and topographical distribution have poor reliability, even when observers are experienced and undergo special training.[1,8]

The most useful development in the classification of CP in recent years has been the development of the Gross Motor Function Classification System (GMFCS).[9] The GMFCS is a five-level ordinal grading system based on the assessment of self-initiated movement with emphasis on function during sitting, standing and walking. Distinctions between different levels are based on functional limitations, the need for walking aids or wheeled mobility and quality of movement (Table 1). Unlike the classification of motor type and topography, the GMFCS has been shown to be a valid, reliable, stable and clinically relevant method for the classification and prediction of motor function in children with CP between the ages of 2 and 12 years.[9,10]

The main aims of this study were to determine the distribution of motor impairment in a large population cohort of young people with CP with respect to the motor type, topographic distribution and gross motor function according to the GMFCS, and to explore the relationships between the three classification methods. The determination of the spectrum of functional limitation that characterizes the group of children with CP is

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Accepted for publication 21 March 2005.

Table 1 Gross Motor Function Classification System (GMFCS) levels, for children aged 6–12 years[9]

GMFCS level I. Children walk indoors and outdoors, and climb stairs without limitation. Children perform gross motor skills including running and jumping but speed, balance and coordination are impaired.
GMFCS level II. Children walk indoors and outdoors, and climb stairs holding onto a railing but experience limitations walking on uneven surfaces and inclines and walking in crowds or confined spaces. Children have at best only minimal ability to perform gross motor skills such as running and jumping.
GMFCS level III. Children walk indoors or outdoors on a level surface with an assistive mobility device. Children may climb stairs holding onto a railing. Children may propel a wheelchair manually or are transported when travelling for long distances or outdoors on uneven terrain.
GMFCS level IV. Children may continue to walk for short distances on a walker or rely more on wheeled mobility at home, school and in the community. Children may achieve self-mobility using a power wheelchair.
GMFCS level V. Physical impairments restrict voluntary control of movement and the ability to maintain antigravity head and trunk postures. All areas of motor function are limited. Children have no means of independent mobility and are transported.

essential to enable planning for resource allocation and to facilitate studies relating to aetiology, prevention or prognosis.[1]

A secondary aim of this study was to compare our study cohort with recently published cohorts that have included motor type, topographical distribution and GMFCS levels. Gorter *et al*. reported the relationships between limb distribution (topographical distribution) motor impairment and functional classification in 657 children, aged from 1 to 13 years at study onset, from Ontario, Canada.[11] Although not based on a CP register, this is a large community-based sample from the originators of the GMFCS. The study by Nordmark *et al*. is smaller ($n = 167$), but is population-based and examines gross motor function, type of motor impairment and topography of children with CP within the public health system of southern Sweden.[12] The time frame of this Swedish study (birth years 1990–1993) is the same as the current study.

METHODS

The Victorian Cerebral Palsy Register (VCPR) was used to identify an inception cohort of children with a confirmed diagnosis of CP and born between January 1990 and December 1992 in the State of Victoria, Australia. These 3 years were chosen to give a large population cohort of children from a period of high-case ascertainment for the register and to allow sufficient follow-up time to determine the true extent of motor impairment and musculoskeletal issues.[1,3]

The VCPR is administered in accordance with state and institutional legislation governing privacy and ethical standards, and attempts to include all children with CP born in the State of Victoria since 1970. Ascertainment of cases is based on a standard definition of CP.[2] Multiple sources of ascertainment are used, including records of patients who access a variety of services at the major paediatric hospitals in Victoria, paediatricians and parent or consumer groups. Motor type, topographical distribution and severity are entered into the VCPR at the time of case ascertainment and confirmed at age 5 years, as is the practice with several other established CP registers.[1] The GMFCS level has recently been added to the VCPR dataset, and is now included in the routine assessment of children in developmental and orthopaedic clinics and in the gait laboratory.

For this study, a cohort of children was selected from the VCPR using the following inclusion criteria:

1. Birth date between January 1990 and December 1992, inclusive;
2. Adequate clinical records to allow determination of functional level according to the GMFCS, between the ages of 6 and 12 years; and
3. Survival to age 6 years.

Children were grouped according to motor type, topographic pattern and GMFCS level. Motor types were classified as spastic, dyskinetic, mixed, ataxic and hypotonic, as defined in Table 2.[5–7] Spastic patterns were further classified according to topographical distribution as hemiplegia (unilateral involvement), diplegia (bilateral involvement, with the lower limbs more affected than the upper limbs) and quadriplegia (bilateral involvement with the upper limbs more or equally involved). Motor type and topographical distribution, as entered on the VCPR, were checked with the child's case records and any discrepancies were resolved by discussion with the child's developmental paediatrician, neurologist or physiotherapist. The motor types were also cross-checked with records from the gait laboratory, where available, and discrepancies were resolved by a full review of clinical records and gait laboratory video tapes and data files.

Children were also classified according to the GMFCS for ages 6 to 12 years. Although the majority of children had already been assigned a GMFCS level independent of this study, multiple records were checked for agreement and discrepancies were resolved by discussion with the child's physiotherapist and developmental paediatrician. Children without an assigned GMFCS level were classified by at least two of the study authors after discussion with the child's community and/or hospital-based physiotherapist.

Stata 8.0 (Stata 2003) was used for the statistical analysis. To explore the relationships between motor type and GMFCS, and topographic distribution and GMFCS, ordered logistic regression was used to calculate odds ratios (OR) and 95% confidence intervals (95% CI) that summarize the propensity for one motor

Table 2 Definitions of motor types

Spasticity	Hypertonia in which resistance to externally imposed movement increases with increasing speed and varies with direction of movement and/or rises rapidly above a threshold speed.
Dyskinesia	Involuntary, sustained or intermittent muscle contractions causing twisting and repetitive movements, abnormal postures or both.
Mixed motor types	Clinical features of more than one type, usually spastic and dyskinetic.
Ataxia	Abnormal pattern of posture and/or movement with loss of orderly muscle coordination so that movements are performed with abnormal force, rhythm or accuracy.
Hypotonia	Abnormally low tone, in the trunk and limbs that must be distinguished from weakness.

Table 3 The distribution of motor types and topographical distribution in 323 children with cerebral palsy, born in Victoria between 1990 and 1992. For each motor type/topographic pattern, the frequency and percentage are shown within each GMFCS level

Motor type/topographic pattern	GMFCS					Total cohort
	I *n* (%)	II *n* (%)	III *n* (%)	IV *n* (%)	V *n* (%)	*n* (%)
Hypertonia						
Spastic						
Hemiplegia	79 (81)	17 (17)	1 (1)	0 (0)	1 (1)	98 (30)
Diplegia	26 (33)	26 (33)	22 (28)	4 (5)	0 (0)	78 (24)
Quadriplegia	1 (1)	3 (3)	13 (13)	36 (35)	50 (48)	103 (32)
Dyskinesia	0 (0)	2 (40)	1 (20)	2 (40)	0 (0)	5 (2)
Mixed	1 (5)	3 (14)	5 (24)	7 (33)	5 (24)	21 (7)
Ataxia	4 (44)	2 (22)	3 (33)	0 (0)	0 (0)	9 (3)
Hypotonia	3 (33)	0 (0)	1 (11)	3 (33)	2 (22)	9 (3)
Total cohort	114 (35)	53 (16)	46 (14)	52 (16)	58 (18)	323 (100)

type or topographical distribution to score higher up the GMFCS scale than a reference group. Proportions of children in different groups were compared between studies using chi-squared tests, and shifts up the GMFCS scale were summarized using OR from an ordered logistic regression model.

RESULTS

There were 374 children on the register with a birth date between January 1990 and December 1992. Nine of these children who died before the age of 6 years were excluded from our analysis. A further 26 children were excluded due to inadequate records and 16 more were lost to follow-up. This left 323 children for analysis, 86% of the cohort. The distributions of motor type, topographic pattern and GMFCS level are shown in Table 3. Eighty-six per cent of children had a spastic motor type, and were relatively evenly distributed between spastic hemiplegia, diplegia and quadriplegia. The other groups with hypertonia (dyskinetic, mixed) accounted for 8% of cases while the remaining motor types (ataxic, hypotonic) represented 6% of the cohort. With respect to the GMFCS classification, level I was the most common at 35%. The remaining cases were distributed fairly equally from levels II to V, in the range of 14–18% per level.

Within the spastic group, differences in motor function among the three topography groups were extremely clear-cut. Compared to children with a hemiplegic distribution, children with diplegia were more severe on the GMFCS scale (OR = 9.3, 95% CI: 4.7, 18.0). Children with spastic quadriplegia had the lowest levels of function, being significantly higher on the GMFCS scale than those with hemiplegia (OR = 341, 95% CI: 246, 1800) and diplegia (OR = 72, 95% CI: 30, 172) (Fig. 1). In comparison with the total group with spasticity, children with both dyskinesia (OR = 1.6, 95% CI: 0.4, 6.2) and hypotonia (OR = 1.8, 95% CI: 0.5, 6.0) appeared to have slightly higher levels of function on the GMFCS scale, but the numbers of children in these groups were too small for a conclusive finding. Children classified as having a mixed type of motor disorder were significantly more severe on the GMFCS scale than children with spasticity (OR = 2.8, 95% CI: 1.3, 5.8) or ataxia (OR = 5.3, 95% CI: 1.4, 20.1).

The exception to these general relationships between topographical distribution and GMFCS was one child with spastic hemiplegia who was graded as GMFCS level V and three children with spastic quadriplegia classified as GMFCS levels I and II. The child with spastic hemiplegia who was graded as GMFCS level V had a hemispherectomy for intractable epilepsy, and was non-ambulant because of a mixture of severe cognitive, behavioural and motor deficits. The children graded as GMFCS levels I and II who had spastic quadriplegia were noted to have

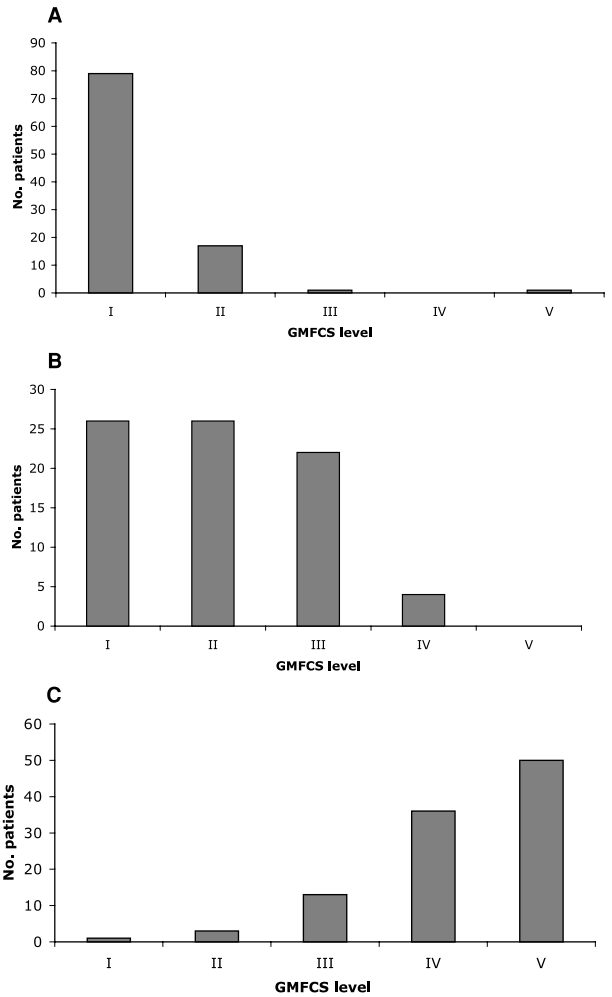

Fig. 1 GMFCS level for 279 children with spastic motor type from a cohort of cerebral palsy cases in Victoria born between 1990 and 1992. (A) GMFCS for children with spastic hemiplegia; (B) GMFCS for children with spastic diplegia; (C) GMFCS for children with spastic quadraplegia.

Table 4 Motor type, topographical distribution and GMFCS levels in children with cerebral palsy in Victoria, Australia; Ontario, Canada; and Southern Sweden

GMFCS	I		II		III		IV		V		Total
Victoria	114	35.3%	53	16.4%	46	14.2%	52	16.1%	58	18.0%	323
Ontario	183	27.9%	80	12.2%	122	18.6%	137	20.9%	135	20.5%	657
Sweden	68	40.7%	31	18.6%	23	13.8%	19	11.4%	26	15.6%	167

Motor type	Spastic		Dyskinetic		Mixed		Ataxic		Hypotonic		Total
Victoria	279	86.4%	5	1.5%	21	6.5%	9	2.8%	9	2.8%	323
Ontario	500	78.2%	39	6.1%	58	9.1%	16	2.5%	26	4.1%	639
Sweden	NA										

Topography	Hemiplegia		Diplegia		Quadriplegia		Total
Victoria	98	35.1%	78	28.0%	103	36.9%	279
Ontario	98	15.3%	217	33.9%	325	50.8%	640
Sweden	50	40.0%	58	46.4%	17	13.6%	125

NA, not applicable.

bilateral upper motor neuron signs in both the upper and lower limbs, principally hyper-reflexia and upgoing plantar responses. However, these three children had good selective motor control, good strength and functioned at a very high level.

Comparison with two other cohorts

The GMFCS levels, motor types and topographical distributions for the three cohorts are shown in Table 4. The Swedish study, like ours, was a cohort derived from a population-based registry. There was a shift in topography towards four limb involvement in our cohort relative to the Swedish cohort (OR = 1.8, 95% CI: 1.2, 2.7, $P = 0.002$), but the distributions of GMFCS were similar (OR = 0.76, 95% CI: 0.54, 1.07). It was not possible to compare motor types as the Swedish study followed a different classification system.

In the Canadian study, a random sample of eligible children was selected that was large enough to ensure at least 60 children in each GMFCS level. There were significant differences in topography compared to both Sweden and Victoria (χ^2 (d.f. = 4) = 94.2, $P < 0.001$); and across the motor types (χ^2 (d.f. = 4) = 14.3, $P = 0.007$). In particular, the Victorian cohort had a higher percentage of spastic motor types (86%) than the Ontario cohort (78%) and a lower percentage of dyskinesia (1.5% vs 6.1%). Compared to the cohort from Ontario, the Victorian cohort (OR = 0.71, 95% CI: 0.56, 0.91) and the Swedish cohort (OR = 0.55, 95% CI: 0.40, 0.74) were both relatively less severe on the GMFCS.

DISCUSSION

The widespread adoption of the definition of CP proposed by Bax in 1964 has resulted in the grouping together of large numbers of children with a wide variety of movement disorders, topographical distributions and functional abilities under the 'cerebral palsy umbrella'.[3,13] The marked variation in functional abilities of children sharing the common diagnosis of CP has led to repeated efforts to classify children into meaningful clinical syndromes. In terms of motor type and topographical distribution, these efforts have had limited success, and this makes it difficult to make meaningful time and geographical comparisons.[1–3,8,9]

By contrast, the use of the GMFCS now facilitates a reliable means of classification of the severity of the motor disorder. The distribution of children throughout all GMFCS levels confirms the wide range of function and disability in a typical population sample of children with CP. GMFCS level I children are completely independent, they do not use aids, and they usually have mild spastic hemiplegia. By contrast, children in GMFCS level V have no independent mobility and often have severe spastic dyskinesia in a quadriplegic distribution. It is of note also, that children with spastic hemiplegia will usually be in levels I and II, children with spastic diplegia will be in levels II, III and IV and those with quadriplegia will be in levels III, IV and V.

This study highlights some of the problems with the commonly used motor type and topography classifications. In collecting data for the VCPR, where multiple sources of information were accessed for any one child, discrepancies were frequently encountered in the identification of the most prominent motor type. Indeed, when the cases were reviewed for this study, common misclassifications were found based on the definitions presented in Table 2. It became clear from comparing the VCPR with gait laboratory records that the five children (1.5%) registered as having dyskinesia all had pure athetoid CP, but a significant proportion of those classified as having a spastic motor type had some element of dyskinesia. For example, many children considered to have spastic hemiplegia, were later found to have a spastic pattern in the lower limb and a dyskinetic pattern in the upper limb, when they presented for upper limb interventions. Many adolescents with spastic quadriplegia were found to have dyskinesia in the upper limbs. These findings are of more than taxonomic importance. Dyskinesia may respond unpredictably to surgery designed for spasticity but favourably to medication.[5–7]

In addition to both random and systematic classification errors, the method used to ascertain cases has the potential to seriously bias the results, particularly if some groups are under-ascertained relative to others. The birth prevalence rate of CP for this cohort is 1.98/1000 live births and this is comparable to the rate of 2.08/1000 determined through a pooling of European CP registers.[14] Despite an overall high level of case ascertainment, it is still possible that children functioning at GMFCS level I may be under-ascertained by the VCPR, as these children are the least likely to access specialist services. In contrast, children classified as GMFCS level IV or V have significant functional disability and associated comorbidities that necessitate early clinical assessment, and practically ensure their identification through the VCPR. Despite being included on the VCPR, children who

died before the age of 6 years were excluded from this study, and this may have resulted in an underestimation of GMFCS V. Children in GMFCS levels II, III and IV develop significant gait deviations, which prompt evaluation by a developmental paediatrician, orthopaedic surgeon or by referral to the gait laboratory. These children are more likely to be reliably ascertained by the register.

The comparison with two other cohorts demonstrates some of these classification and ascertainment issues. Classification by topographical distribution showed the greatest variability between the three studies. The Canadian study had a higher incidence of quadriplegia, reflecting the referral-based nature of their population cohort. The Swedish study[12] classified topography quite differently, defining spastic quadriplegia as 'massive total motor disability, the upper limbs at least as affected as the lower limbs and non-ambulant'.

The poor reliability of topographical classification is due to the inconsistency of distinctions between 'severe diplegia' and quadriplegia, and between asymmetrical hemi-syndromes and bilateral CP. The use of body diagrams to describe the topographical distribution in CP, based on the presence or absence of spasticity or limb involvement, appears promising but requires validation. Efforts have also been made by the European group to standardize topographical classifications and motor type.[15]

The Victorian CP register is already an invaluable research tool but clinicians need further education about the need to better define, assess and report dyskinesia when classifying motor type. We need to further examine the possibility that significant changes in motor type are occurring between early childhood, when children are classified and entered on the register, and later in childhood when some of the critical decisions regarding spasticity management and gait correction surgery must be made.[6,16]

In conclusion, the term 'cerebral palsy', despite all its shortcomings, is worth retaining, although it might be better to use the term 'cerebral palsies' to describe a CP phenotype that encompasses the enormous variability in motor type, topography and gross motor function.[3] We think that the best way of classifying children with CP is a combination of motor type, topography and gross motor function, according to the GMFCS. Although the motor type and topographical classifications lack reliability, they are familiar, widely used and clinically significant. In the meantime, the GMFCS is a simple, intuitive and reliable tool to classify gross motor function in children with CP. It has modest correlations with measures of speech, hearing, vision and cognition and strong correlations with certain musculoskeletal problems.[14] These will be examined in future studies. The GMFCS is recommended as a tool for the improvement of communication between health professionals involved in the care of children with CP.

ACKNOWLEDGEMENTS

We acknowledge the contribution of Chris Blackburn, physiotherapist, from the Monash Medical Centre, and Mary Sheedy, from the Department of Orthopaedics at the Royal Children's Hospital.

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