

# CEREBRAL PALSY IN WESTERN SWEDEN

## Epidemiology and function

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*"There's nothing you can do that can't be done"*

*Lennon/McCartney*



## Abstract

**Aims:** To investigate the prevalence and aetiology of cerebral palsy (CP), describe and analyse motor function and accompanying impairments, apply a new classification of unilateral and bilateral CP and describe prevalence, aetiology, function and growth in dyskinetic and bilateral spastic CP.

**Material and methods:** In a population-based study in western Sweden, the prevalence and aetiology of CP were analysed in children born in 1995-1998. Gross and fine motor function, accompanying impairments and, in the case of dyskinetic and bilateral spastic CP, neurology and growth were recorded in the 1991-1998 birth cohort. For dyskinetic CP, neuroimaging and perinatal factors were reviewed. The prevalence and severity of motor impairment in the birth-year period 1959-1998 were analysed.

**Results:** The prevalence of CP was 1.92 per 1,000 live births. Spastic hemiplegia, diplegia and tetraplegia accounted for 38%, 35% and 6%, dyskinetic CP for 15% and ataxia for 6% respectively. The aetiology in children born at term was considered to be prenatal in 38%, peri/neonatal in 35% and unclassifiable in 27%. In children born preterm, it was 17%, 49% and 33% respectively. Gross Motor Classification System (GMFCS) levels were distributed at level I in 32%, level II in 29%, level III in 8%, level IV in 15% and level V in 16%. Learning disability was present in 40%, epilepsy in 33% and severe visual impairment in 19%. The severity of the motor impairment correlated to the presence of accompanying impairments and, in children born at term, to the presence of adverse peri/neonatal events. The prevalence of dyskinetic CP was 0.27 per 1,000 live births. The majority were dystonic, 79% were unable to walk and spasticity was present in 69%. Learning disability was present in 73%, epilepsy in 63% and 79% had anarthria. In the children born near term or at term, peri/neonatal adverse events had been present in 81%. The motor impairment was most severe in this group. Neuroimaging revealed isolated late third-trimester lesions in 56% and a combination of early and late third-trimester lesions in 16%. The prevalence of bilateral spastic CP was 0.69 per 1,000 live births. After 1975, children born preterm dominated. A severe motor impairment was found in 46% of the children born at term and in 33% of those born preterm. The GMFCS correlated with the severity of spasticity and deviation in growth.

**Conclusions:** The prevalence of CP continued to decrease, especially in those born preterm. Hemiplegia was the most common CP type, due to a decrease in preterm diplegia. CP type and motor function combined was an indicator of the total impairment load. Gestational age at birth and peri/neonatal morbidity provided prognostic information. Classification into unilateral and bilateral spastic CP combined with GMFCS level added structure to the CP classification. Dyskinetic CP was dominated by term-born, appropriate for gestational age children, with severe disabilities and underweight at follow-up. Peri/neonatal adverse events were common. The prevalence of bilateral spastic CP had decreased, parallel to a decrease in the severity of motor impairment. Spasticity correlated with motor function.

**Key words:** cerebral palsy, prevalence, aetiology, motor function, dyskinetic, bilateral spastic, growth



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## List of publications

- I Himmelmann K, Hagberg G, Beckung E, Hagberg B, Uvebrant P.  
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*Acta Paediatr* 2005;94:287-294.
- II Himmelmann K, Beckung E, Hagberg G, Uvebrant P.  
Gross and fine motor function and accompanying impairments in cerebral palsy.  
*Dev Med Child Neurol* 2006;49:417-423.
- III Himmelmann K, Hagberg G, Wiklund LM, Uvebrant P. Dyskinetic cerebral palsy – a population-based study of children born in 1991-1998.  
*Dev Med Child Neurol* 2006; *accepted for publication*.
- IV Himmelmann K, Beckung E, Hagberg G, Uvebrant P. Bilateral spastic cerebral palsy – epidemiology, function and growth.  
*Submitted for publication*.



## Abbreviations

AAC	Augmentative and alternative communication
AGA	Appropriate for gestational age
ACOG	American College of Obstetricians and Gynecologists
BFMF	Bimanual Fine Motor Function
BMI	Body mass index
CHQ	Child Health Questionnaire
CNS	Central nervous system
CP	Cerebral palsy
CT	Computed tomography
df	Degrees of freedom
GA	Gestational age
GMFCS	Gross Motor Function Classification System
GMFM	Gross Motor Function Measure
HIE	Hypoxic-ischemic encephalopathy
ICF	International Classification of Functioning, Disability and Health
ICIDH	International Classification of Impairments, Disabilities and Handicaps
IQ	Intelligence quotient
LGA	Large for gestational age
MACS	Manual Ability Classification System
MRI	Magnetic resonance imaging
NE	Neonatal encephalopathy
ns	Non-significant
PNM	Perinatal mortality
QoL	Quality of life
SCPE	Surveillance of Cerebral Palsy in Europe
SD	Standard deviation
SGA	Small for gestational age
WHO	World Health Organization



## Introduction

### Definition

Cerebral palsy (CP) is the most common cause of motor disability in childhood. It comprises a group of conditions, heterogeneous in cause and manifestations, described and classified over the years by authors such as Little (Little 1862), McNutt (McNutt 1885), Freud (Freud 1897), Gowers (Gowers 1888), Osler (Osler 1889) to the present day. To date, the definition formulated by Mutch et al. has been used in Sweden (Mutch et al. 1992). The definition of CP as a pure motor impairment has been challenged (Shapiro 2004). The various brain lesions causing the motor dysfunction often also impair sensation, vision, cognition, communication and behaviour and may cause epilepsy. A definition of CP including the naming of some accompanying impairments was put forward by the participants at an international workshop on the definition and classification of CP, held in Washington in July 2004 (Bax et al. 2005):

*Cerebral palsy (CP) describes a group of 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 disturbance of sensation, cognition, communication, perception, and/or behaviour, and/or by a seizure disorder.*

In this definition more emphasis than before is placed on the accompanying impairments. The focus on accompanying impairments is growing, as we learn more about perception and specific learning difficulties and how to assess them. The new definition is a result of this more comprehensive way of looking at CP. However, the classification rests firmly upon type and distribution of motor impairment. Common to all definitions is the insult to an immature brain leading to motor impairment, the non-progressiveness and the multi-factorial aetiology. It is said that “there are greater differences between the brain of a 28-week gestation infant and that of a 36-week infant than there are between the brain of a three-month old baby and an adult” (Pape and Wigglesworth 1979). In spite of this, a motor impairment caused by any insult to the developing brain can eventually be affiliated to the identifiable pattern that is the foundation of CP classification.

### Classification

In Sweden and other countries, the classification by Hagberg has been used for more than four decades (Hagberg et al. 2001). Consensus in classification, regarding both the CP concept and its different syndromes (SCPE 2002, Ashwal et al. 2004) is important to avoid misconceptions about the aetiology and severity of disability. Population-based series are necessary for international comparisons and epidemiological studies of trends (Bax 2004). Only in well-defined popula-

tions, can aspects such as prevention, the prevalence of perception and behavioural problems, as well as participation (Beckung and Hagberg 2002) and the provision of health care, be studied. For the sake of European cooperation, a common classification has been put forward by fourteen centres throughout Europe, the Surveillance of Cerebral Palsy in Europe, SCPE (SCPE 2000, 2002). This classification is in accordance with the recently proposed definition and classification of CP (Bax et al. 2005). It includes the concept of unilateral and bilateral spastic CP, and emphasizes diagnosis by the dominant syndrome. The term bilateral spastic CP serves as an alternative to the often-confusing terms such as diplegia, tetraplegia, double hemiplegia and quadriplegia, with varying definitions between countries. The former gives us means not only to describe the group, but also to compare populations, evaluate changes and predict needs in these particular groups.

### **Prevalence**

The overall prevalence of CP may appear to have been fairly stable over the years, but major changes have occurred in the various subgroups (Hagberg et al. 1975a, 1975b, 1982, 1984, 1989a, 1993, 1996, 2001). The advances in maternal health care, obstetric and neonatal care have resulted in changes in the prevalence of CP in children born preterm (Hagberg et al. 1989b, 2000), a decrease in children born at low gestational age in some countries (Surman et al. 2003) and an increase in others (Blair and Watson 2006). At the same time, more fragile children of all gestational ages survive than before, with chronic morbidity of various kinds, such as lung disease and neurodevelopmental impairment other than CP (Cooke 2006, Msall 2006). Spastic CP types are by far the most common, constituting 80-85% of the CP panorama (Hagberg et al. 2001, Nordmark et al. 2001). The prevalence of dyskinetic CP varies the most between countries, probably due to the fact that dyskinetic children with additional spasticity have been diagnosed as spastic in many countries (Hagberg et al. 2001, SCPE 2002).

### **Function and health**

In western Sweden, population-based CP studies have monitored prevalence and aetiology as well as functional aspects of CP and its accompanying impairments for many consecutive years (Hagberg et al. 2001). The development of new reliable and valid measures of function has facilitated the recording of gross motor function (Palisano et al. 1997, 2000) and, in accordance with an emerging interest (Bax 2004), a classification of fine motor function, Bimanual Fine Motor Function (BFMF) was recently added (Beckung and Hagberg 2002). Aspects of function and the prerequisite for activity and participation have attracted growing interest in recent years. The International Classification of Functioning, Disability and Health (ICF) supplies one framework, within which the different aspects of health, or absence of health, can be described (WHO 2001), as opposed to its predecessor International Classification of Impairments, Disabilities and Handicaps (ICIDH) which was more disease oriented (WHO 1980, Baxter 2004). Bartlett and Palisano have proposed another multivariate model to determine

motor outcome in cerebral palsy, including the characteristics of the child, such as motor and accompanying impairments, but also family ecology (Bartlett and Palisano 2000).

The Gross Motor Function Classification System (GMFCS) has been widely accepted as a classification of motor function and has been adapted to different age bands (Palisano et al. 1997, 2000, Rosenbaum et al. 2000). Its stability over time has been investigated and reported (Wood and Rosenbaum 2000, Palisano et al. 2006). A further sub-classification of CP based on limb distribution has been proposed, but Gorter et al. showed that this did not add prognostic value to classification with the GMFCS (Gorter et al. 2004). The GMFCS correlates with the former ICIDH handicap code (WHO 1980, Beckung and Hagberg 2000). The Manual Ability Classification System (MACS) will further facilitate surveys of hand function in daily activities (Eliasson et al. 2006). Both the BFMF and the MACS are divided into five levels, to correspond to the GMFCS.

The Gross Motor Function Measure (GMFM) tests five dimensions of gross motor function, from performance lying down to jumping and running, related to the performance of a healthy five-year-old child (Russell et al. 1989, Palisano et al. 1997). The inter- and intra-rater reliability is good (Nordmark et al. 1997). It is suggested that GMFM scores plateau at six to seven years of age (Bartlett and Palisano 2000) and consequently the test has less value in older children with good motor function. Spasticity, a hallmark of the majority of children with CP, contributes to the motor impairment as well as the muscle weakness (Engsberg et al. 1998, 2000), lack of selective motor control and balance (Rose et al. 2002, Liao and Hwang 2003, Woollacott and Shumway-Cook 2005), and accompanying impairments, such as visual impairment and learning disability (Jacobsson et al. 1996, Stiers et al. 2002, Aylward 2002).

The importance of strength has been described, as well as its correlation with gross motor function (Damiano and Abel 1998, Damiano et al. 2002a, Ross and Engsberg 2002). Predictors of future walking are of interest for the planning of interventions (Bleck 1975, Campos da Paz et al. 1994, Sala and Grant 1995, Fazzi 2000).

### **Accompanying impairments**

In more than half the children with CP, there are accompanying impairments, which may override the motor impairment in some. Epilepsy (Carlsson et al. 2003), learning disability of varying degree and profile (Fennell and Dikel 2001, Pueyo et al. 2003), severe visual (Jacobson and Dutton 2000, Jan et al. 2001, Jacobson et al. 2002, Stiers and Vandebussche 2004) and hearing impairments may be possible to detect at an early age, while more subtle sensory and cognitive problems eventually become apparent at school age (Jacobson et al. 1996, Barnett et al. 2002, Shenker et al. 2005, Lindström 2006). Screening for these conditions should be part of the evaluation of a child with CP (Russman and Ashwal 2004). There is emerging evidence of behavioural disorders (Elgen et al. 2002, Fedrizzi et al. 2003, Bax et al. 2005).

## **Aetiology**

When CP was first described in 1862, the aetiology was attributed to the perinatal period (Little 1862). This was objected to by Freud (Freud 1887). An attempt to classify CP by aetiology was made by Sachs and Peterson (Sachs and Peterson 1890). Since then, there has been a vigorous debate. Moreover, the aetiological profiles differ considerably between developed and developing countries (Blair and Watson 2006). Many prenatal aetiological factors have been discussed (Blair and Stanley 1992, Badawi et al. 1998a, Nelson and Grether 1999, Nelson 2003) such as infection and inflammation (Grether and Nelson 1997, Jacobsson et al. 2002, Grether et al. 2003), environment and genetics (Evrard et al. 1997), multiple pregnancy (Williams et al. 1996) and intrauterine growth restriction (Jarvis et al. 2003, Topp et al. 2004, Glinianaia et al. 2006). Factors of less importance in developed countries today are mainly rhesus immunisation with subsequent hyperbilirubinemia and congenital rubella. However, perinatal factors such as asphyxia, hypoxic-ischemic encephalopathy (HIE), or neonatal encephalopathy (NE) (Thornberg et al. 1995, Badawi et al. 1998b, Moster et al. 2001, Milsom et al. 2002, Thorngren-Jerneck et al. 2002, Pierrat et al. 2005, Badawi et al. 2005) have remained the subject of interest. Comparisons between studies have been complicated by the various definitions of HIE and NE that are in use (Dilenge et al. 2001). The criteria for considering perinatal events as the aetiology of CP put forward by the American College of Obstetricians and Gynecologists in collaboration with the American Academy of Pediatrics are strict and only applicable to children with a gestational age of 34 weeks or more (ACOG 2003).

In 2000, Stanley et al. introduced causal pathways as a new aetiological model for cerebral palsy, in an attempt to “elucidate cause while maintaining an open mind about the several possible models of causation”, i.e. a sequence of events culminating in CP (Stanley et al. 2000).

In the study of cerebral palsy in western Sweden, the criteria for referring to prenatal, peri/neonatal and postnatal aetiological period have been basically the same, throughout the four decades (Hagberg et al. 1996). During this time, new methods of investigation, mainly MRI, have revealed more of the underlying causes and timing of events (Barkovich et al. 2001, Cowan et al. 2003, Flodmark et al. 2003, Krägeloh-Mann 2004, Kwong et al. 2004, Ricci et al. 2006). It has been proposed that neonatal MRI findings predict neurological outcome in preterm infants with periventricular leucomalacia (Sie et al. 2005) and in term children with lesions in the internal capsule (Rutherford et al. 1998).

## **Consequences**

### ***Growth disturbance***

Another subject that has attracted attention is growth in children with CP. The cause of growth disturbance in this group is the subject of debate, and probably multi-factorial. Reilly et al. found oral motor dysfunction in the majority of children with CP in a community survey (Reilly et al. 1996). Malnutrition (Stallings et al. 1993), growth hormone deficiency (Coniglio et al. 1996) and other non-nutritional causes (Stevenson et al. 1994, 1995) have been discussed. The pattern



of sexual maturation is altered in CP (Worley et al. 2000), contemporary with a growth plateau rather than a growth spurt in adolescence (Stevenson et al. 2006). Osteopenia has been reported in the majority of children with CP performing at GMFCS levels III-V in a population-based sample by Henderson et al. (Henderson et al. 2002). Feeding difficulties, anticonvulsants and nutritional status were contributory factors. Fractures were not uncommon.

### ***Pain***

Pain is particularly difficult to assess in severely impaired children, and it is therefore often underestimated. However, it is often also overlooked in more able children. Several reports have shown frequent and chronic pain in children with CP (Breau et al. 2003, Houlihan et al. 2004, Engel et al. 2005). Prejudice regarding pain in children with cognitive impairment among both the caregivers and health care professionals may affect the children's care (Breau et al. 2003a, 2003b). Pain affects the opportunity to participate and has social and educational consequences (Houlihan et al. 2004).

### ***Quality of life***

Quality of life (QoL) in moderate and severe CP was worse compared with that in children without CP in a study by Liptak et al., using the Child Health Questionnaire (CHQ) which is one of the most common instruments (Liptak et al. 2001). Among parents of children with CP, the CHQ revealed a reduced QoL related to the severity of CP expressed as the GMFCS level (Vargus-Adams 2005). The ratings appear to be stable over time (Bjornson and McLaughlin 2001, Vargus-Adams 2006). An increasing number of QoL instruments, including a condition-specific measure of QoL in CP (Waters et al. 2005) are being developed. The diversity of measures, often focusing on difficulties rather than positive aspects, makes it difficult to compare results (Davis et al. 2006).

### ***Adult life with CP***

Adults with CP are sometimes overlooked in modern health care, when it comes to addressing problems related to spasticity (Andersson and Mattsson 2001) and pain (Engel et al. 2003) as well as the even less obvious accompanying problems of cognitive impairment and epilepsy (Michelsen et al. 2005). Adults with CP may face social isolation and unemployment. In a survey of young adults with CP, Ng et al. found significantly less contact with specialists and therapists after leaving school (Ng et al. 2003). Fatigue is frequently reported by adults with CP (Jahnsen et al. 2003).

### ***Survival***

CP may be associated with full life expectancy. However, survival is related to severity of motor impairment and accompanying impairments (Hutton and Pharoah 2002, 2006, Hemming et al. 2006). Blair found that intellectual disability was the strongest predictor of mortality in a survey of birth years 1958-1994, while severe motor impairment primarily increased early mortality (Blair et al. 2001).



## **Aims**

The aims of the study were

To investigate the prevalence and aetiology of CP in the birth-year period from 1995 to 1998, as a continuation of the Panorama of Cerebral Palsy Study

To describe and analyse gross and fine motor function and accompanying neurological impairments in the birth-year period from 1991 to 1998 and apply the SCPE classification of unilateral and bilateral CP and severe CP

To describe the prevalence, aetiology including neuroimaging findings, motor impairment, accompanying impairments and growth in dyskinetic CP

To depict changes in the prevalence and severity of bilateral spastic CP over a 40-year period and to characterise the group born in 1991-1998 with respect to gross motor function, spasticity and growth



## Material

### Study area

The present study was performed in western Sweden. The study area comprised the counties of Västra Götaland, Jönköping and Halland, with a total population of 2.1 million inhabitants and a slightly positive net migration (Figure 1). The County of Uppsala was included in 1959-1967.

### The birth-year period 1959-1998

In the birth-year period from 1959 to 1998 there were 901,928 live births in the study area. Of these, 1,770 were diagnosed with cerebral palsy. The severity of motor impairment was known in 1,683 subjects.

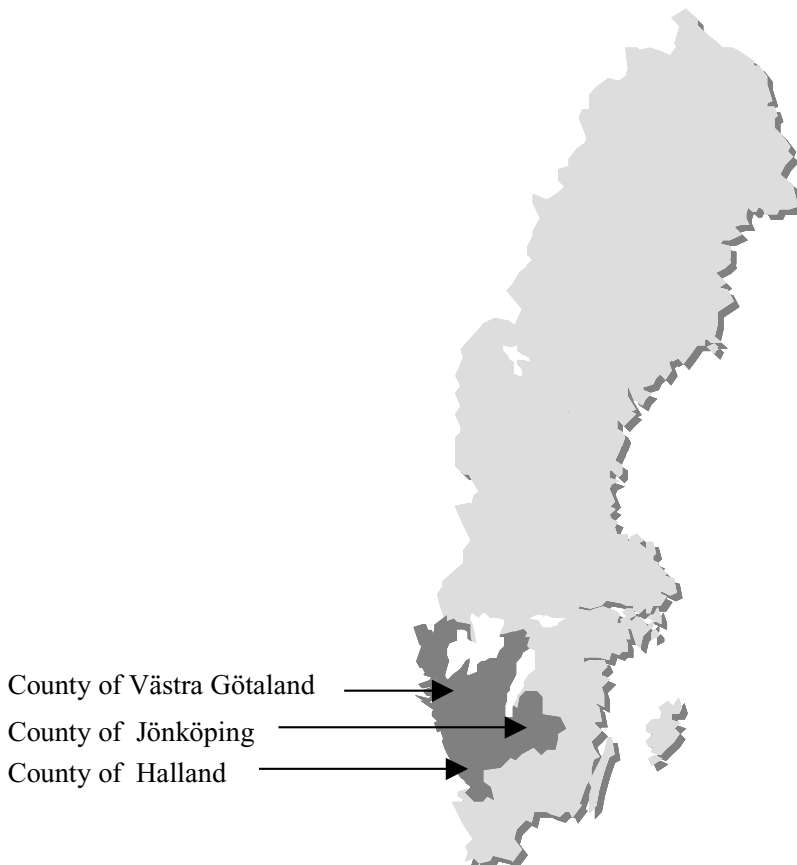


Figure 1. Study area in the western part of Sweden.

## **The birth-year period 1991-1998**

### *I. The changing panorama of cerebral palsy in Sweden. IX. Prevalence and origin in the birth-year period 1995-1998*

In 1995-1998, there were 88,371 live births in the study area. There were 170 children with a confirmed diagnosis of CP at the age four to eight years, eight of whom were postneonatally derived. Of the remaining 162, 92 were born at term and 70 preterm (Figure 2).

### *II. Gross and fine motor function and accompanying impairments in cerebral palsy*

Of the 411 children with CP in the study area born 1991-1998, the GMFCS level was known in 367. The remaining 44 children did not differ from those included in the study in terms of CP type or gestational age. The BFMF level was known in 345 and a complete record of accompanying impairments was at hand in 353 children (Figure 2). Evaluation was done at four to eight years of age.

### *III. Dyskinetic cerebral palsy - a population-based study of children born in 1991-1998*

Fifty-five of the 411 children in the study area born 1991-1998 with CP were identified with dyskinetic CP. Forty-eight (87%) agreed to participate in a further evaluation at the age of five to 13 years; mean age nine years (Figure 2).

### *IV. Bilateral spastic cerebral palsy – prevalence through four decades, motor function and growth*

Of the 411 children in the study area born in 1991-1998 with CP, 167 with a known GMFCS level were identified with bilateral spastic CP, 144 of whom had diplegia and 23 tetraplegia (Figure 2 and 3, Table 1). Follow-up was performed at the age of four to 12 years; mean age seven years.

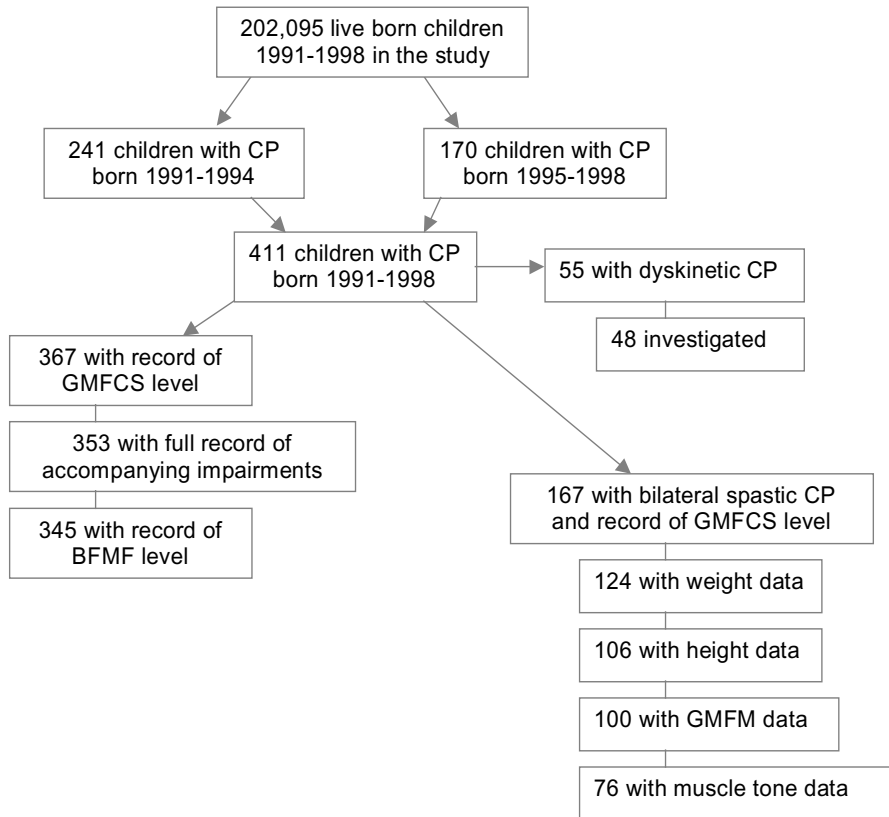


Figure 2. An overview of the material, children born 1991-1998.

Table 1. Available information on GMFM, growth and muscle tone at follow-up in 145 of 167 children (87%) with bilateral spastic CP born 1991-1998.

Additional information	n
Growth <sup>a</sup>	28
Growth + GMFM <sup>b</sup>	21
Growth + GMFM + muscle tone <sup>b</sup>	59
Growth + muscle tone <sup>c</sup>	16
GMFM	21
Total	145

<sup>a</sup> height missing in seven, <sup>b</sup> in five, and <sup>c</sup> in one.

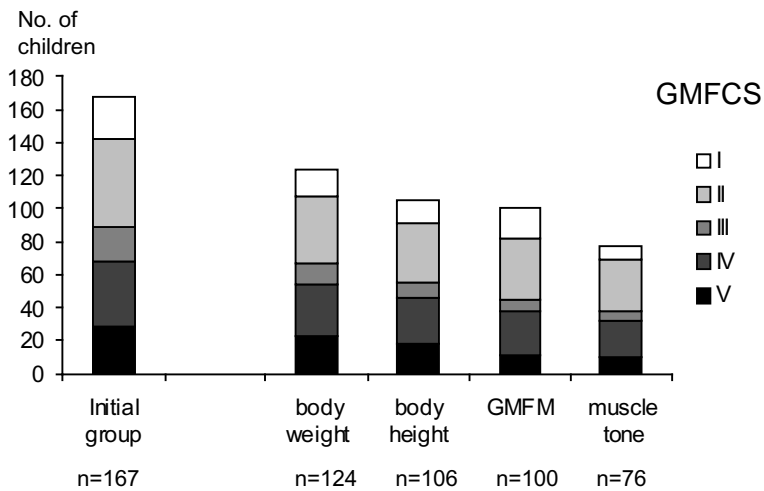


Figure 3. The distribution of Gross Motor Function Classification System (GM-FCS) levels at follow-up, for each variable (children with data on body weight, body height, Gross Motor Function Measure (GMFM) and muscle tone). The subgroups did not differ in GMFCS distribution from the initial group.



## Methods

### Definition

The definition of CP in the present study was that agreed at an international consensus meeting in 1990 (Mutch et al. 1992). According to this definition CP is “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 development”.

### Classification

The Swedish and internationally accepted classification of CP by Hagberg was applied (Mutch et al. 1992). The classification according to SCPE was used in parallel (SCPE 2002) (Table 2).

*Table 2. A comparison of the two CP classifications used in this study (Mutch et al. 1992, SCPE 2002).*

	Hagberg	SCPE
Spastic	Hemiplegia	Unilateral spastic CP
	Tetraplegia	Bilateral spastic CP
	Diplegia	
Ataxic	- diplegia - congenital (simple)	Ataxia
Dyskinetic	- Mainly dystonic - Mainly choreoathetotic	- Dystonic - Choreo-athetotic

### Obstetric and birth data

Extremely preterm birth was defined as birth occurring before 28 completed gestational weeks, very preterm at 28-31 weeks, moderately preterm at 32-36 weeks and birth at term at more than 36 weeks, based primarily on ultrasound early in pregnancy. If this information was not available, menstrual data were used.

Prenatal referred to the period of pregnancy until the onset of labour resulting in delivery, perinatal to the period from the onset of labour until the seventh day of life, neonatal to the period up to day 28 and postneonatal to the period from day 29 to two years of age.

Maternal disorders were acute severe illness during pregnancy, e.g. pyelonephritis, or fever of  $> 38.5^{\circ}\text{C}$  at delivery, or pre-existing chronic disorder, e.g. diabetes mellitus, celiac disease, essential hypertension, epilepsy, pharmacological treat-

ment of psychiatric disorder, hypothyroidism and nephropathy (Uvebrant 1988). Small for gestational age (SGA) was defined as a birth weight for gestational age of  $\leq -2SD$ , appropriate for gestational age (AGA) as  $>-2SD$  and  $\leq 2SD$ , and large for gestational age (LGA) as  $> 2SD$  from the mean on a Swedish growth chart (Nicklasson et al. 1991).

### Aetiological period

The aetiological classification was based on given clinical criteria, combined with available neuroimaging information (Hagberg et al. 1996) (Table 3). Ob-

*Table 3. Aetiological criteria according to Hagberg (Hagberg et al. 1996). Items higher on the list taking precedence over those lower on the list with the exception of peri/neonatal intra-cerebral haemorrhage/infarction, neonatal shock and brain oedema.*

<b>Prenatal</b>		
Intrauterine infection		
CNS malformations		
Other prenatal CNS abnormalities		
	For children born at $\geq 34$ weeks of gestation with normal delivery and peri/neonatal period:	periventricular atrophy/ porencephaly
<b>Perinatal/neonatal</b>		
- Most likely	Intracranial haemorrhage/ infarction/neonatal shock/ brain oedema	
	CNS infection and/or sepsis	
	For children born at $\geq 34$ weeks of gestation:	hypoxic-ischemic encephalopathy
	For children born at $< 34$ weeks of gestation:	Periventricular atrophy and/or intracerebral haemorrhage with normal initial ultrasound
- Probable	For children born at $< 34$ weeks of gestation:	Low Apgar score and/ or low pH/mechanical ventilation $> 7$ days or complicated with pneumothorax
<b>Unclassifiable</b>		

stetric and peri/neonatal data, as well as CP type, were derived from medical and habilitation records.

### Neuroimaging

The findings at computed tomography (CT) and/or magnetic resonance imaging (MRI) were classified into six categories; i.e. malformations, periventricular atrophy, cortical/subcortical atrophy, basal-ganglia lesions, other findings and normal. Cerebral/cerebellar malformations were considered in the case of documented neural migration disorders, aplasia/hypoplasia and prenatal cysts. Intracerebral haemorrhage was graded according to Papile et al. (Papile et al. 1978). In a re-evaluation of the MRI and CT of the children with dyskinetic CP, a timetable adapted from Krägeloh-Mann (Barkovich et al. 2001, Krägeloh-Mann 2004) was used (Table 4).

Table 4. Pattern of brain maldevelopments or lesions by the stage of brain development. Adapted from Krägeloh-Mann (Krägeloh-Mann 2004).

1 <sup>st</sup> and 2 <sup>nd</sup> trimester	Maldevelopments
Anencephaly	
<i>Disorders of proliferation</i>	
Microcephalia vera, hemimegalencephaly, cortical dysplasia with balloon cells	
<i>Disorders of migration</i>	
Lissencephaly, pachygyria, heterotopias	
<i>Disorders of organization</i>	
Schizencephaly, polymicrogyria (until week 30)	
Hydranencephaly	
3 <sup>rd</sup> trimester	Lesions
<i>Early/mid 3<sup>rd</sup> trimester</i>	White matter
Intracranial hemorrhage	
Periventricular infarction	
Periventricular leucomalacia	
Multicystic encephalomalacia	
Thromb-embolic lesions	
<i>Late 3<sup>rd</sup> trimester</i>	Gray matter
Basal ganglia/thalamus lesions +/-	
Cortico-subcortical lesions of the central region	
Parasagittal lesions	
Multicystic encephalomalacia	
Thromb-embolic lesions	

## **Hypoxic-ischemic encephalopathy**

HIE was considered in children born at  $\geq 34$  weeks of gestation in the presence of two or more of the following symptoms or signs: (a) Apgar score  $< 5$  at one or five minutes; (b) resuscitation/ subsequent mechanical ventilation and (c) convulsions before day 3 (Krägeloh-Mann et al. 1995). The combination of only resuscitation and a low Apgar score was not considered as HIE. The criteria for birth asphyxia severe enough to cause CP in children born  $\geq 34$  weeks of gestation were chosen from criteria suggested by Stanley et al. (Stanley et al. 2000). They were considered when a series of the following four events was present: (a) intrauterine hypoxia (discoloured amniotic fluid, foetal heart rate during labour of  $< 100$  or  $> 160$  beats per minute, silent pattern or dip 2 pattern on cardiotocography, cord prolapse or placental ablation); (b) Apgar score  $< 5$  at one or five minutes; (c) subsequent mechanical ventilation or convulsions before day 3 and, when performed (d) normal findings at early neuroimaging or evidence of acute cerebral abnormality. Birth asphyxia was considered to have started intrapartum when Apgar scores of 0-6 were documented for longer than five minutes (MacLennan 1999).

## **Risk factors**

The following possible risk factors associated with CP were recorded:

Low Apgar score ( $< 5$  at one or five minutes), multiple birth, maternal acute severe illness or chronic disorder, SGA and LGA.

## **Peri- or neonatal adverse events**

Children without major prenatal anomaly or lesion, who had intracranial haemorrhage/stroke, cerebral infection (viral or bacterial meningitis/meningoencephalitis) or HIE were judged as having had a peri- or neonatal adverse event.

## **Accompanying impairments**

Learning disability was defined as mild in children with an estimated or measured intelligence quotient (IQ) of 50-70 and severe if the IQ was less than 50.

Epilepsy was defined as a diagnosis of active epilepsy at four to eight years of age, requiring medical treatment.

Severe visual impairment was defined as functional blindness or an acuity after correction of refraction errors of  $\leq 0.3$  (20/60) in the better eye.

Severe hearing impairment was defined as the need for a hearing aid or no hearing.

## **Hydrocephalus**

Infantile hydrocephalus was defined as a diagnosis of surgically treated expansive hydrocephalus in the first year of life.

## **Gross and fine motor function**

The criteria for the five levels of the GMFCS and the BFMF, relevant for the ages studied, are described in Table 5 (Rosenbaum et al. 2002, Beckung and Hagberg 2002).

Table 5. Classification of gross and bimanual fine motor function (Rosenbaum et al. 2002, Beckung and Hagberg 2002).

<b>GMFCS</b>	<b>BFMF</b>
Level I	Level I
Walks without restrictions. Limitations in more advanced gross motor skills.	One hand: manipulates without restrictions. The other hand: manipulates with restrictions or limitations in more advanced fine motor skills
Level II	Level II
Walks without restrictions. Limitations walking outdoors and in the community.	a) One hand: manipulates without restrictions. The other hand: only ability to grasp or hold. b) Both hands: limitations in more advanced fine motor skills.
Level III	Level III
Walks with assistive mobility devices, limitations walking outdoors and in the community.	a) One hand: manipulates without restrictions. The other hand: no functional ability. b) One hand: limitations in more advanced fine motor skills. The other hand: only ability to grasp or worse.
Level IV	Level IV
Self-mobility with limitations, children are transported or use power mobility outdoors and in the community.	a) Both hands: only ability to grasp. b) One hand: only ability to hold. The other hand: only ability to hold or worse.
Level V	Level V
Self-mobility is severely limited, even with the use of assistive technology.	Both hands: only the ability to hold or worse.

### **Gross motor function test**

The GMFM is a test of five dimensions of gross motor function (Russell et al. 1989). A total score is obtained by calculating the mean percentage of the points obtained for each dimension. The maximum achievement is that of a healthy five-year-old (Table 6).

### **Muscle tone**

Muscle tone was assessed in selected muscles according to the modified Ashworth scale by Peacock-Staudt (Peacock and Staudt 1991). A score of 1 represents normal muscle tone (Table 7).

Table 6. Gross Motor Function Measure (Russell et al. 1989).

Dimensions	
A	lying and rolling
B	sitting
C	crawling and kneeling
D	standing
E	walking, running and jumping
T	total score

Table 7. Modified Ashworth score according to Peacock and Staudt (Peacock and Staudt 1991).

Score	Grade	Description
0	Hypotonic	Less than normal muscle tone, floppy
1	Normal	No increase in muscle tone
2	Mild	Slight increase in muscle tone, 'catch' in limb movement or minimal resistance to movement through less than half of the range
3	Moderate	Marked increase in muscle tone through most of the range of motion but the passive movement of the affected part is easily performed
4	Severe	Considerable increase in muscle tone, passive movement difficult
5	Extreme	Affected part rigid in flexion or extension

## Dystonia

Video recordings of each child were used to evaluate dystonia in the eyes, mouth, neck, trunk, arms and legs according to the five-grade Barry-Albright Dystonia Scale (Barry et al. 1999). A total dystonia score was calculated as the sum of the six partial scores (Table 8).

Table 8. Barry-Albright Dystonia Scale (Barry et al. 1999).

Dystonia scale	
0	Absence of dystonia
1	Less than 10% of the time and does not interfere with activity
2	Less than 50% of the time and does not interfere with activity
3	More than 50% of the time and/or interfering with activity
4	More than 50% of the time and/or prevents activity

### **Anthropometric measures**

Body weight and height at follow-up were recorded (Albertson-Wikland et al. 1994) and body mass index (BMI) calculated (Karlberg et al. 2001). Children with dyskinetic CP were followed up at a mean age of nine years (range 5-13 years) and those with bilateral spastic CP at a mean age of 7.1 years (range 4-12 years).

### **Acute intrapartum hypoxic event**

The criteria to define an acute intrapartum hypoxic event were first presented by MacLennan representing the International Cerebral Palsy Task Force in 1999 and were further processed by the American College of Obstetricians and Gynecologists in 2003 (MacLennan 1999, ACOG 2003). They are given in Table 9.

*Table 9. ACOG Task Force on Neonatal Encephalopathy and Cerebral Palsy, 2003 (ACOG 2003).*

#### **Essential criteria (must meet all four)**

1. Evidence of a metabolic acidosis in fetal umbilical cord arterial blood obtained at delivery (pH <7.00 and base deficit  $\geq$ 12mmol/l)
2. Early onset of severe or moderate neonatal encephalopathy in infants born at 34 or more weeks of gestation
3. Cerebral palsy of the spastic quadriplegic or dyskinetic type
4. Exclusion of other identifiable etiologies such as trauma, coagulation disorders, infectious conditions, or genetic disorders

#### **Criteria that collectively suggest an intrapartum timing (within close proximity to labor and delivery, eg, 0-48 hours) but are non-specific to asphyxial insults**

5. A sentinel (signal) hypoxic event occurring immediately before or during labor
6. A sudden and sustained fetal bradycardia or the absence of fetal heart rate variability in the presence of persistent, late, or variable decelerations, usually after a hypoxic sentinel event when the pattern was previously normal
7. Apgar scores of 0-3 beyond 5 minutes
8. Onset of multisystem involvement within 72 hours of birth
9. Early imaging study showing evidence of acute nonfocal cerebral abnormality

### **Statistical methods**

The frequency of cerebral palsy is expressed as prevalence per 1,000 live born, based on the agreed view that CP is a permanent impairment and that the reported frequency is the number of children with CP in a specific age span (Stanley et al. 2000). In this study, the prevalence on a specific census date in a defined study area and population has been used. The following statistical tests were used: chi-square test, trend in proportion in analyses of proportions. For small groups, Fisher's test was used. For correlations: Spearman's rho, Kendall's tau b

and Pearson's correlation coefficient were used. A paired t-test was used for comparison of paired continuous data. The level of significance was set at  $p < 0.05$ . All the statistics and p-values have been interpreted in a descriptive manner.



## CP history of western Sweden

### The changing panorama of cerebral palsy in Sweden study

Western Sweden has one of the longest running CP registers in the world, comprising data from birth year 1954 to 1998. Prevalence, aetiology and accompanying impairments has been registered and reported every four years from a population living in a defined study area. To date nine reports have been published. A large number of other papers have emerged from these data, shedding light on the various CP types, e.g. ataxia (Sanner and Hagberg 1974), dyskinetic CP (Kyllerman et al. 1982), diplegia (Veelken et al. 1983), spastic tetraplegia (Edebol-Tysk et al. 1989) and hemiplegia (Uvebrant 1988). This register is one of the original contributors to the European collaboration, the SCPE.

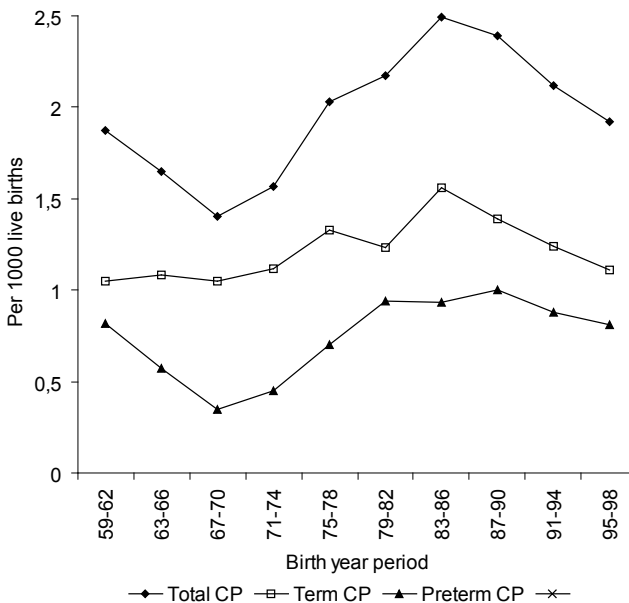


Figure 4. Prevalence of CP during four decades, 1959-1998.

The results from the changing panorama of cerebral palsy in Sweden study presented here are based on data from 1,770 individuals with CP born in the four decades ranging from 1959 to 1998 (Figure 4). In 1,683 (95%) of them, the severity of motor impairment at four to eight years of age was known. The classification was limited to mild (walks without aids), moderate (walks with aids) and severe (wheel-chair dependent). Figure 5 shows the prevalence and severity of the gross motor impairment in the whole CP group born between 1959 and 1998.

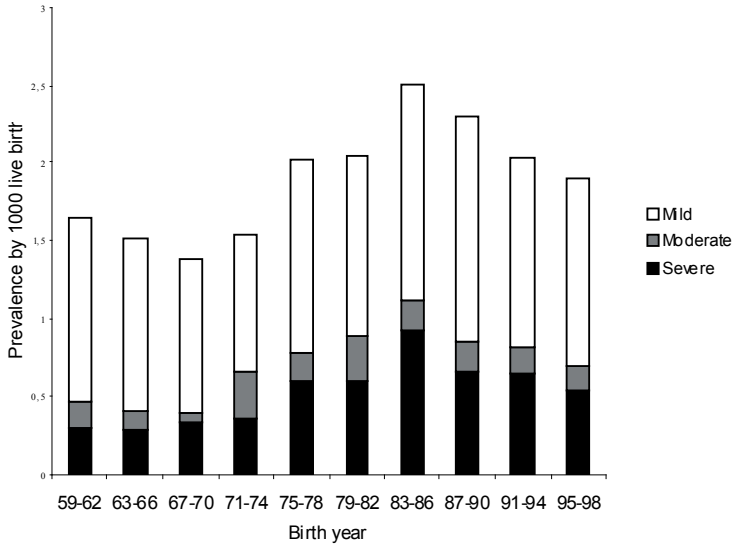


Figure 5. Prevalence and severity of motor impairment in CP 1959-1998, based on data from 1,683 individuals in a population-based study.

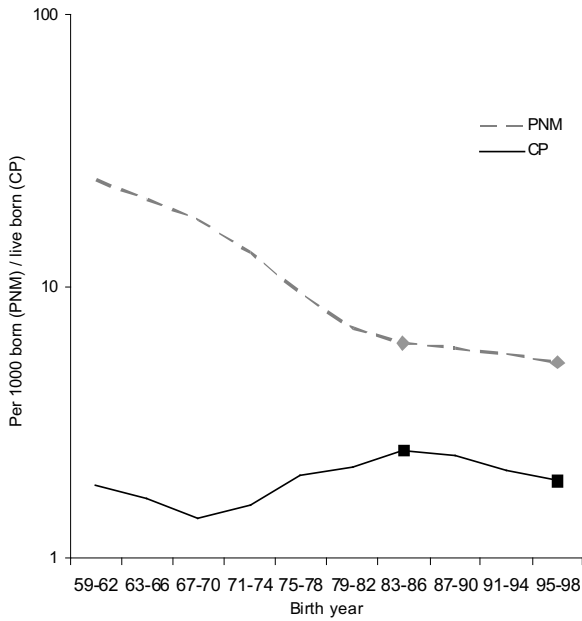


Figure 6. Gains and hazards of modern maternal, perinatal and neonatal care. A comparison between the birth-year periods 1983-1986 and 1995-1998.

Since the mid-1980s the prevalence of CP has decreased significantly ( $p < 0.01$ ). A contemporary decrease in perinatal mortality from 6.21 in the birth-year cohort 1983-1986 to 5.27 per 1,000 in 1995-1998 has taken place. During the same period, the crude CP prevalence decreased from 2.49 to 1.92 per 1000 live births, which added to the survivors without CP (Table 10).

Had there been the same perinatal mortality and CP prevalence in the late 1990s as in the mid-1980s, 83 children would not have survived, and 50 more would have been diagnosed with CP.

*Table 10. A comparison of outcome in the birth-year period 1983-1986 and 1995-1998.*

All CP	1983-1986	1995-1998	Live born 1995-1998 n=88,371
Perinatal mortality (per 1,000 born)	6.21	5.27	
CP prevalence (per 1,000 live born)	2.49	1.92	
Gains in saved lives		0.94	83
Additional CP		-0.37	-50
Gain in saved lives without CP			133



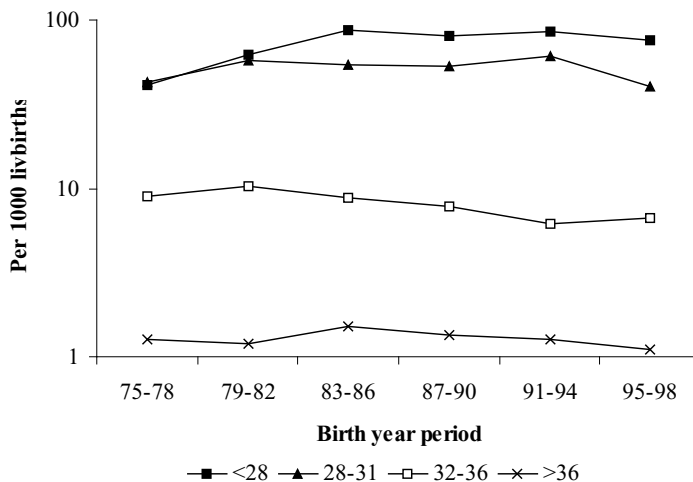
## Results

### ***I. The changing panorama of cerebral palsy in Sweden. IX. Prevalence and origin in the birth-year period 1995-1998***

The present series comprised 170 children with CP born in 1995-1998. The crude mean prevalence of CP was 1.92 per 1,000 live births, 0.81 per 1,000 for preterm CP and 1.11 per 1,000 for term CP. The decreasing prevalence in both the term and preterm group since the 1980s reached statistical significance in the late 1990s ( $p < 0.01$ ).

Excluding eight postneonataly derived cases, the gestational age-specific prevalences were 77 per 1,000 for children born before 28 weeks of gestation, 40 for children born at 28-31 weeks, seven for children born at 32-36 weeks and 1.1 for children born after 36 weeks of gestation (Figure 7). From the period 1983-1986, the previous rising trend in the extremely preterm group had stabilised. A decreasing trend ( $p = 0.11$ ) had occurred in the very preterm group between the birth-year period 1991-1994 and 1995-1998. In children born moderately preterm and at term, the decrease continued and was now also statistically significant in the term group ( $p < 0.05$ ).

The birth-weight-specific prevalence was 82.0 per 1,000 for a birth weight of  $< 1,000$  g, 54.4 for a birth weight of 1,000-1,499 g, 6.7 per 1,000 for a birth weight of 1,500-2,499 g and 1.2 per 1,000 for a birth weight of 2,500 g or more. Eight children (4.9%) were born SGA and six (3.7%) were born LGA.



*Figure 7. Gestational age-specific prevalence of CP. Postneonatal cases are excluded.*

Of the 162 children, 96 of whom were boys, sixteen children (10%) were born extremely preterm, 23 (14%) very preterm, 31 moderately preterm (19%) and 92 (57%) at term. Sixteen children (10%) had a birth weight of < 1,000 g, 22 (14%) a birth weight of 1,000-1,499 g, 22 (14%) a birth weight of 1,500-2,499 g and 102 (63%) a birth weight of 2,500 g or more. Multiple births resulted in 23 children (14%) with CP.

Spastic hemiplegia, diplegia and tetraplegia accounted for 38%, 35% and 6% respectively, dyskinetic CP for 15% and ataxia for 6%.

Compared with earlier periods, hemiplegia outnumbered diplegia for the first time (Figure 8). This was mainly explained by the decrease in diplegia associated with post-haemorrhagic hydrocephalus in the very preterm group. The increasing trend for dyskinetic CP continued and was now 0.28 per 1,000 or 15% of all CP.

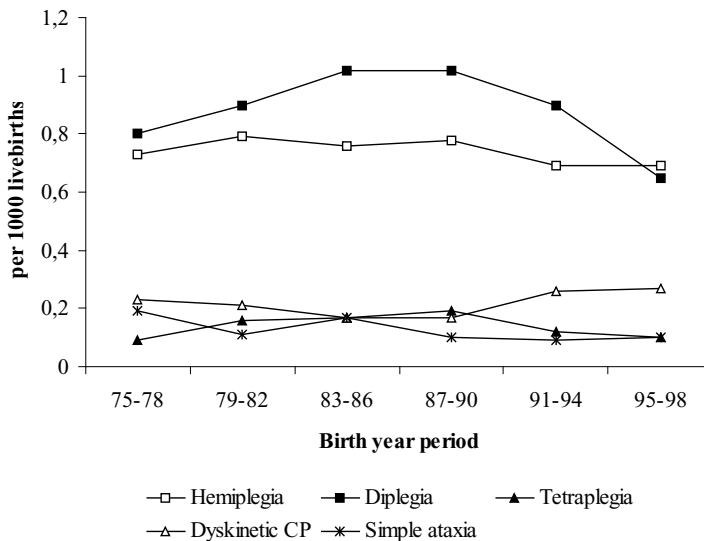


Figure 8. Prevalence of CP by CP type. Postneonatal cases are excluded.

### Neuroimaging

The findings in 129 CT and MRI examinations are presented by gestational age in Table 11. The most frequent main finding was periventricular atrophy, present in 40% of the prenatal group, 25% of the peri/neonatal and 30% of the unclassifiable group. Cortical/subcortical atrophy was present in 19% and basal-ganglia lesions in 13%.

### Risk factors

Low Apgar scores were associated with a high risk of CP in children born at term. Compared with all the children born at term in the study area in 1995-1998, the risk of developing CP increased 36-fold for an Apgar score of < 5 at five min-

Table 11. Findings in CT and MRI by gestational age in 129 children with CP.

	Gestational age				Total
	< 28 wk	28-31 wk	32-36 wk	> 36 wk	
CNS malformations		1	3	11	15
Periventricular atrophy	6	9	12	15	42
Cortical/subcortical lesions	5	3	3	14	25
Basal-ganglia lesions	1 <sup>a</sup>		2	14	17
Other findings		1		9	10
Normal		4		16	20
Total	12	18	20	79	129

<sup>a</sup> This child had periventricular atrophy and basal ganglia lesions, the latter deriving from severe asphyxia at 44 weeks of post-conceptual age.

Table 12. Type of CP according to Apgar scores in 89 children born at term with CP.

	Apgar at 1 min Median (range)	Apgar at 5 min Median (range)	Apgar at 10 min Median (range)
Hemiplegia, n=41	9 (2-10)	10 (4-10)	10 (8-10)
Spastic/ataxic diplegia, n=17	8 (1-9)	9 (3-10)	10 (3-10)
Tetraplegia, n=3	9 (1-10)	10 (3-10)	10 (6-10)
Dyskinetic CP, n=20	2 (0-9)	4 (0-10)	5 (2-10)
Simple ataxia, n=8	9 (6-10)	10 (7-10)	10 (9-10)

utes. In children born at term, CT/MRI findings were strongly correlated to CP type as well as Apgar scores at one, five and 10 minutes ( $p < 0.001$ ). Dyskinetic CP was associated with the lowest Apgar scores, compared with the other CP subtypes (Table 12). Children with basal-ganglia lesions had the lowest Apgar scores at five minutes, while children with CNS malformations and periventricular atrophy had the highest scores (Table 13).

Multiple birth was present in 21 of 152 children (14%), fourteen of whom were born preterm. Two twin sisters born at 25 weeks of gestation both developed CP. Three co-twins had died prenatally. Seven of nine children born after assisted fertilisation were twins.

Maternal disorder was present in 38 mothers (25%). Fever at delivery and maternal diabetes were the most frequent events, present in 10 and nine mothers respectively.

Table 13. Main findings at neuroimaging (CT and/or MRI) according to Apgar scores in 89 children with cerebral palsy born at term.

	Apgar at 1 min Median (range)	Apgar at 5 min Median (range)	Apgar at 10 min Median (range)
CNS malformations, n=11	9 (8-10)	10 (9-10)	10 (9-10)
Periventricular atrophy, n=15	9 (4-10)	10 (8-10)	10 (10-10)
Cortical/subcortical lesions, n=14	6.5 (1-9)	8.5 (3-10)	9.5 (4-10)
Basal-ganglia lesions, n=14	2 (0-10)	3.5 (1-10)	6.5 (2-10)
Other, n=9	7.5 (1-9)	9 (3-10)	9 (7-10)
Normal findings, n=16	9 (0-9)	9 (0-10)	10 (2-10)
Not done, n=10	9 (3-10)	10 (3-10)	10 (4-10)

Eight children were born SGA, three of whom were twins born preterm. The remaining five all suffered from placental insufficiency. Six children were born LGA.

If these risk factors were applied to the 45 children with an unknown cause of CP, an additional 19 cases, all but one born preterm, could probably be allocated to an aetiological time period. Eleven would have been confined to the prenatal and four to the peri/neonatal period. In four cases, there were both pre- and perinatal risk factors.

### Aetiology

The origin of CP in children born at term was considered to be prenatal in 38%, peri/neonatal in 35% and unclassifiable in 27%. In children born preterm the origin was considered to be prenatal in 17%, peri/neonatal in 49% and unclassifiable in 33% (Table 14, Figure 9).

## II. Gross and fine motor function and accompanying impairments in cerebral palsy

### Gross motor function

The gross motor function expressed as the GMFCS was at level I in 116 (32%), level II in 108 (29%), level III in 30 (8%), level IV in 56 (15%) and level V in 57 (16%) of the 367 children. The distribution differed significantly between CP types ( $p < 0.001$ ) (Table 15).

In children with spastic hemiplegia, 127 of 134 (95%) were classified at GMFCS levels I-II, mainly level I. In children with spastic diplegia, 78 of 144 (54%) were classified as GMFCS levels I- II, with most (38%) at level II, whereas 45 (31%)



performed at levels IV and V. Of the 23 children with spastic tetraplegia two performed at GMFCS level IV and the remaining 21 at level V. Among the children with dyskinetic CP, 26 of 51 (50%) were assigned to level V. All but one child with ataxia were affiliated to GMFCS levels I-II, with 12 of 14 at level II (Figure 10).

Table 14. Distribution of aetiology by gestational age in 152 children born 1995-1998.

Gestational age	Preterm			Term	Total
	< 28wk	28-31wk	32-36wk	> 36 wk	
Prenatal	0	3	8	34	45
Intrauterine infection	0	0	1	2	3
CNS malformation	0	1	4	11	16
Other prenatal CNS abnormalities	0	2	0	3	5
For children born at $\geq 34$ weeks of gestation with normal delivery and peri/neonatal period: periventricular atrophy/porencephaly	0	0	3	18	21
Perinatal/neonatal	14	8	9	31	62
<i>Most likely</i>					
Intracranial haemorrhage/infarction/ neonatal shock/brain oedema	8	7	5	6 <sup>a</sup>	26
CNS infection and/or sepsis	1	0	1	3	5
For children born at $\geq 34$ weeks of gestation: Hypoxic-ischemic encephalopathy	0	0	1	22	23
For children born at < 34 weeks of gestation: Periventricular atrophy and/or periventricular haemorrhage with normal initial ultrasound	3	1	2	0	6
<i>Probable</i>					
For children born at < 34 weeks of gestation: Low Apgar and/or low pH/mechanical ventilation > 7 days or complicated by pneumothorax	2	0	0	0	2
Unclassifiable	1	11	9	24	45
Total	15	22	26	89	152

<sup>a</sup> HIE present in three cases.

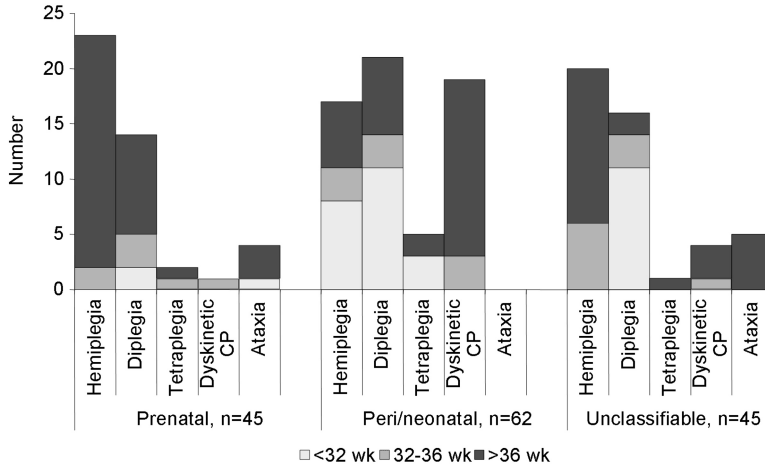


Figure 9. Distribution of aetiology by type of CP and gestational age group in 152 children with CP born in 1995-98. Postneonatal cases and children without parental consent have been excluded.

The proportion of children with the mildest motor impairment, i.e. GMFCS I, increased with gestational age ( $p < 0.001$ ). In children born at term, the distribution of GMFCS levels was significantly associated with the occurrence of adverse peri/neonatal events i.e. intracranial haemorrhage/stroke, cerebral infection and HIE. Forty-two percent of these children were able to walk independently (GMFCS levels I-II) as opposed to 76% without these complications ( $p < 0.001$ ). This difference originated principally from the subgroup of children with a HIE, of whom only 38% were able to walk without aid.

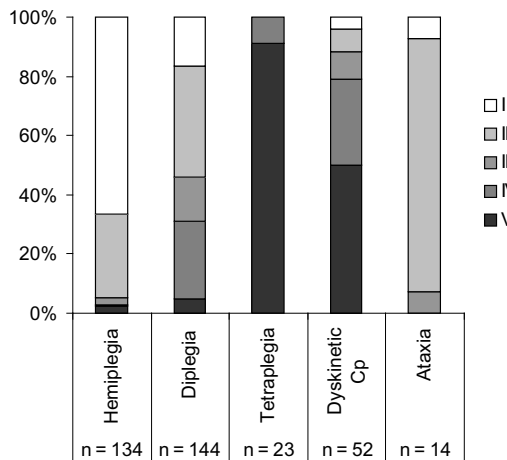


Figure 10. Distribution of Gross Motor Function Classification System levels by CP type in 367 children with CP.

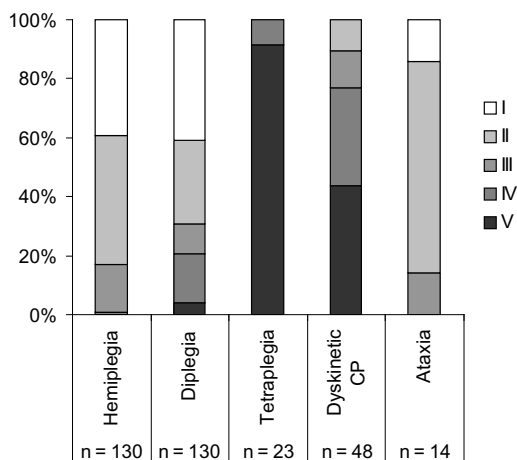


Figure 11. Distribution of Bimanual Fine Motor Function levels by CP type in 345 children with CP.

### Bimanual fine motor function

The distribution of BFMF levels in 345 children is shown in Figure 11; 106 (31%) performed at level I, 109 (32%) at level II, 42 (12%) at level III, 41 (12%) at level IV and 47 (14%) at level V. The distribution differed significantly between CP types ( $p < 0.001$ ) and largely followed the pattern of the GMFCS (Figure 11). The BFMF level was consistent with that of GMFCS in 57% (Table 15). Hemiplegia dominating in the arm with the BFMF level exceeding the GMFCS level was found in 42%. There was a correlation between peri/neonatal compromise and BFMF levels ( $p < 0.001$ ).

### Accompanying impairments

Learning disability was present in 40%, epilepsy in 33%, severe visual impairment in 19% and hearing impairment in 2%. Hydrocephalus was present in 7%. The percentage of children with accompanying impairments increased significantly with GMFCS levels ( $p < 0.001$ ), as shown in Figure 12. In children with motor function at GMFCS level I, 91 of 115 (79%) had no accompanying impairment, in contrast to three of 54 (6%) of those at GMFCS level V. In children performing at GMFCS level V, 48 of 54 (89%) had two or more accompanying impairments.

CP types characterised by milder impairments of gross motor function, i.e. hemiplegia and ataxia, had fewer accompanying impairments. At least one impairment was present in 31% of children with hemiplegia, in 59% of those with diplegia and in all the children with tetraplegia. Learning disability was milder in children with hemiplegia and diplegia than in the other CP types. Epilepsy was most frequent in tetraplegia followed by dyskinetic CP (Table 16). Hydrocephalus was predominantly found in children with diplegia (11%) and tetraplegia (15%).

Table 15. Association between the Gross Motor Function Classification System (GMFCS) and Bimanual Fine Motor Function (BFMF) in 367 children with CP. Children with unknown BFMF levels (n=22) are shown by GMFCS level.

		B F M F					BFMF unknown	Total
CP type		I	II	III	IV	V		
G M F C S	I Hemiplegia	41	39	5	-	-	4	89
	Diplegia	21	2	-	-	-	1	24
	Dyskinetic CP	-	2	-	-	-	-	2
	Ataxia	-	1	-	-	-	-	1
	Subtotal	<b>62</b>	<b>44</b>	<b>5</b>	-	-	<b>5</b>	<b>116</b>
	II Hemiplegia	9	18	11	-	-	-	38
	Diplegia	26	22	2	-	-	4	54
	Dyskinetic CP	-	3	1	-	-	-	4
	Ataxia	2	9	1	-	-	-	12
	Subtotal	<b>37</b>	<b>52</b>	<b>15</b>	-	-	<b>4</b>	<b>108</b>
	III Hemiplegia	-	-	3	-	-	-	3
	Diplegia	5	7	4	-	-	5	21
	Dyskinetic CP	-	-	4	1	-	-	5
	Ataxia	-	-	1	-	-	-	1
	Subtotal	<b>5</b>	<b>7</b>	<b>12</b>	<b>1</b>	-	<b>5</b>	<b>30</b>
	IV Hemiplegia	1	-	-	-	-	-	1
	Diplegia	1	6	7	19	2	3	38
	Tetraplegia	-	-	-	-1	2	-	2
	Dyskinetic CP	-	-	-	2	2	1	15
	Subtotal	<b>2</b>	<b>6</b>	<b>7</b>	<b>31</b>	<b>6</b>	<b>4</b>	<b>56</b>
	V Hemiplegia	-	-	2	1	-	-	3
	Diplegia	-	-	-	4	3	-	7
	Tetraplegia	-	-	-	2	19	-	21
	Dyskinetic CP	-	-	1	2	19	4	26
	Subtotal	-	-	<b>3</b>	<b>9</b>	<b>41</b>	<b>4</b>	<b>57</b>
<b>Total</b>		<b>106</b>	<b>109</b>	<b>42</b>	<b>41</b>	<b>47</b>	<b>22</b>	<b>367</b>

Differences between CP types were significant. Levels II-III and IV-V were combined.  
GMFCS: Chi-square=232.4, df=8, p<0.0001. BFMF: Chi-square=195.5, df=8, (p < 0.0001).

The children born before 28 weeks of gestation had the highest percentage of impairments. In this group, 19 of 37 (51%) had a learning disability with an IQ of <70 and 12 (32%) had epilepsy, 10 (27%) were severely visually impaired and nine (24%) had hydrocephalus. Children born at term were less frequently affected in every aspect except for epilepsy (Table 17).

Accompanying impairments by adverse peri/neonatal events (intracranial haemorrhage/stroke, cerebral infection or hypoxic-ischaemic encephalopathy) in children born at term are shown in Table 18. Accompanying impairments were present in 69% of those who had suffered peri/neonatal adverse events and 41% had two or more impairments. In children without peri/neonatal compromise, 36% had accompanying impairments and 21% of these had two or more.

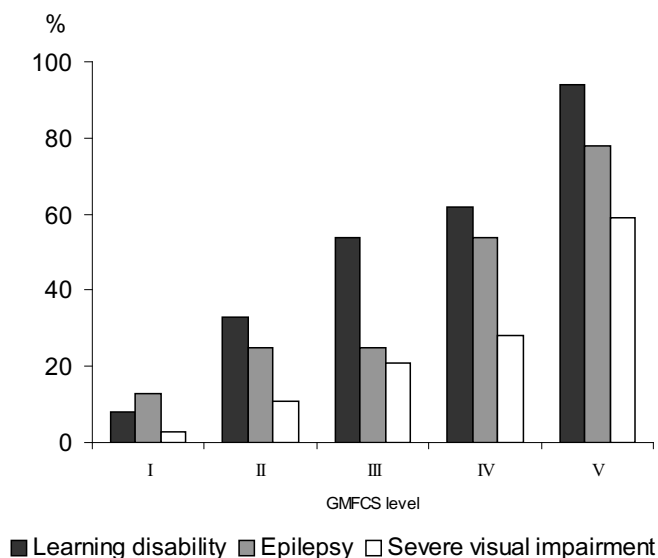


Figure 12. Percentage of learning disability, epilepsy and severe visual impairment by Gross Motor Function Classification System (GMFCS) levels in 353 children with CP. The percentage increased significantly with GMFCS levels.  $\text{Chi-square}_{\text{trend}}$  for learning disability=127.14,  $df=1$ ,  $p<0.0001$ ;  $\text{Chi-square}_{\text{trend}}$  for epilepsy=77.99,  $df=1$ ,  $p<0.0001$ ;  $\text{Chi-square}_{\text{trend}}$  for severe visual impairment=73.59,  $df=1$ , ( $p < 0.0001$ ).

Table 16. The distribution of accompanying impairments by CP type in 353 children with CP. The difference between CP-types was significant.  $\text{Chi-square}=83.6$ ,  $df=8$ ,  $p<0.001$ .

	Hemi- plegia n = 134 n (%)	Diple- gia n = 137 n (%)	Tetra- plegia n = 20 n (%)	Dyski- netic CP n = 50 n (%)	Ataxia n = 12 n (%)	Total n = 353 n (%)
Learning disability	22 (17)	64(49)	20(100)	30(62)	5 (50)	141(40)
Mild	15	37	0	9	5	66
Severe	7	27	20	21	0	75
Epilepsy	30 (23)	43 (34)	18 (87)	26 (52)	1 (7)	118 (33)
Severe visual impairment	10 (8)	26 (21)	17 (83)	12 (27)	1 (8)	66 (19)
<b>No. of accompany- ing impairments</b>						
0	92 (66)	56 (41)	0	14 (28)	7 (50)	169 (48)
1	23 (17)	36 (26)	0	11 (22)	3 (36)	73 (21)
$\geq 2$	19 (15)	45 (32)	20 (100)	25 (50)	2 (14)	111 (31)

## Bilateral spastic CP

Within the new concept of bilateral spastic CP, diplegia and tetraplegia are regarded an entity. The tetraplegic children have the most severe motor impairment, and neuroimpairment load. There was a considerable difference between GMFCS levels IV and V in terms of accompanying impairments. At GMFCS level IV 21% had no accompanying impairment as compared to 3% at level V

Table 17. Accompanying impairments by gestational age (GA) completed weeks in 353 children with CP. The proportion of children with accompanying impairments increased significantly with lower gestational age.  $\text{Chi-square}_{\text{trend}} 5.47, df=1, p<0.05$ .

	GA <28 w n = 37 n (%)	GA 28-31 w n = 55 n (%)	GA 32-36 w n = 55 n (%)	GA >36 w n = 206 n (%)	Total n = 353 n (%)
Learning disability	19 (51)	25 (44)	24 (43)	73(37)	141(40)
Mild	11	14	10	31	66
Severe	8	11	14	42	75
Epilepsy	12 (32)	16 (28)	17 (30)	73 (36)	118 (33)
Severe visual impairment	10 (27)	17 (30)	13 (20)	26 (14)	66 (19)
No. of accompanying impairments					
0	13 (35)	23 (42)	24 (44)	109 (53)	169 (48)
1	9 (24)	13 (24)	12 (22)	39 (19)	73 (21)
≥2	15 (41)	19 (35)	19 (35)	58 (28)	111 (31)

Table 18. Distribution of accompanying impairments in relation to peri/neonatal adverse events. Differences between groups: <sup>a</sup> $\text{Chi-square}=13.44, df=1, p=0.0002$ ; <sup>b</sup> $\text{Chi-square}=11.28, df=1, p=0.0008$ ; <sup>c</sup> $\text{Chi-square}=3.25, df=1, p=0.07$ ; <sup>d</sup> $\text{Chi-square}=13.44, df=2, p<0.0001$ .

	Adverse peri/neonatal events		
	Present n = 71 n (%)	Not present n = 136 n (%)	Total n = 207 n (%)
Learning disability <sup>a</sup>	37 (52)	36 (26)	73 (35)
Mild	13	18	31
Severe	24	18	42
Epilepsy <sup>b</sup>	36 (51)	37 (27)	73 (35)
Severe visual impairment <sup>c</sup>	13 (18)	13 (10)	26 (13)
No. of accompanying impairments <sup>d</sup>			
0	22 (31)	87 (64)	109 (53)
1	20 (28)	20 (15)	40 (19)
≥2	29 (41)	29 (21)	58 (28)

( $p < 0.001$ ). At GMFCS level IV 31% had no learning disability, while the corresponding figure at level V was 3% ( $p < 0.001$ ).

### Trends in unilateral spastic CP

In the birth-year period 1995-1998 more very preterm children with unilateral spastic CP were without additional impairments as compared to the birth-year period 1991-1994. The right side was more often affected than the left in both cohorts (53%). In the children born in 1995-1998, 49% had a poorer bimanual fine motor than gross motor function, compared with 36% of those born in 1991-1994. There was a decrease in the percentage of children at GMFCS level I and a corresponding rise in GMFCS level II between the groups, as seen in Table 19. A decrease in BFMF level I was found, as well as a larger proportion of BFMF level II and III (Table 20).

### Trends in extremely preterm children with CP

During the 1990s the perinatal mortality in children born before 28 completed weeks of gestation decreased from 379 per 1,000 in the birth years 1991-1994 to 285 per 1,000 in the birth years 1995-1998. At the same time the prevalence of CP in this gestational age group decreased from 86 in 1991-1994 to 77 per 1,000 live births in 1995-1998. Within the group of extremely preterm children the percentage of children born before 26 completed weeks had increased from 17% in 1991-1994 to 50% in 1995-1998 ( $p = 0.04$ ). Bilateral spastic CP had decreased in favour of unilateral spastic CP ( $p = 0.028$ ).

Table 19. GMFCS levels in unilateral spastic CP.

GMFCS	1991-1994 n=70 (%)	1995-1998 n=63 (%)	
I	54 (77)	35 (55)	p=0.007
II	14 (20)	25 (40)	p=0.011
III	0 (0)	3 (5)	
IV	1 (1.4)	0 (0)	
V	1 (1.4)	0 (0)	

Table 20. BFMF levels in unilateral spastic CP

BFMF	1991-1994 n=66 (%)	1995-1998 n=64 (%)	
I	33 (50)	18 (29)	p=0.013
II	27 (41)	30 (48)	ns
III	6 (9)	15 (24)	p=0.021
IV	0	1	
V	0	0	

### III. Dyskinetic cerebral palsy - a population-based study of children born in 1991-1998

#### Prevalence

During the period 1991-1998, there were 202,095 live births in the study region. Fifty-five children were identified as having dyskinetic CP, corresponding to a prevalence of 0.27 per 1,000 live-born children. In 48 children (87%), a clinical investigation was performed at a mean age of nine years (range 5-13 years). The live birth prevalence trend of dyskinetic CP from 1959 to 1998 is shown in Figure 13. No significant change over the 40-year period was found.

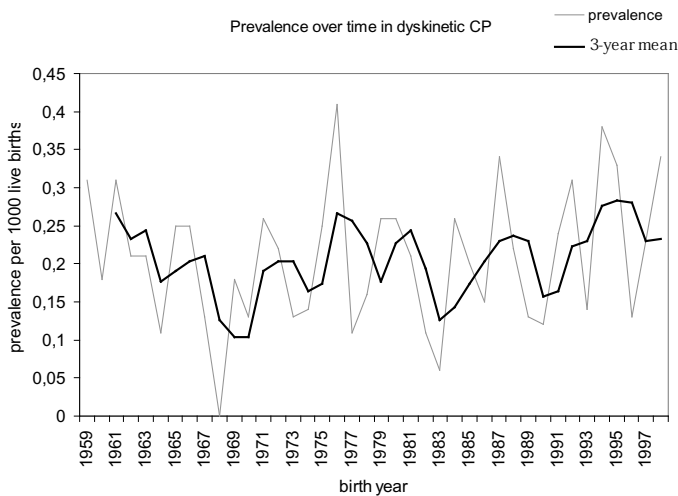


Figure 13. Live birth prevalence of dyskinetic CP 1959-1998 in western Sweden.

#### Clinical picture

##### Neurology

Thirty-seven children (81%) were affiliated to the dystonic group and the remaining nine to the choreo-athetotic group. Primitive reflexes, predominantly asymmetric tonic neck reflex, but also symmetric tonic neck reflex, grip reflex and labyrinth reflex, were present in 43 of the 48 children (90%). The five children without primitive reflexes all had mild motor impairments and four were able to walk unaided and one with aids. Dystonia in the arms was more severe or as severe as in the legs in 40 of the 48 children (83%). Dystonia in the mouth was present in 34 children (71%) and five had detectable eye dystonia (10%). Five had a total dystonia score of 0-4, whereas the dystonia score was 5-9 in seven, 15-19 in 16 and 20-24 in eight respectively. The distribution of dystonia score by GM-FCS level is shown in Figure 14.

Signs of spasticity, i.e. exaggerated tendon reflexes and increased velocity-dependent increase in muscle tone, were present in 33 of the 48 children (69%). In the



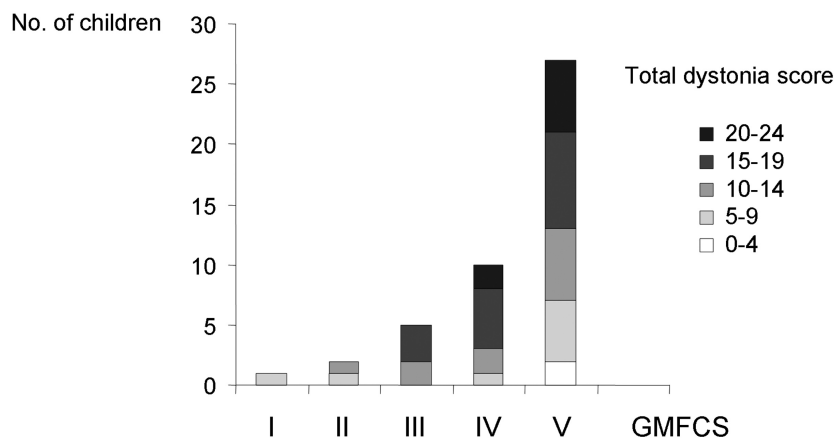


Figure 14. Distribution of total dystonia score by GMFCS level in 48 children with dyskinetic CP.

children with spasticity, extended reflex zones were found in 25, contra-lateral reflex spreading in 15 and clonus in 24. According to the Ashworth score, spasticity was more pronounced in the legs in 21 children, equally affecting the arms in ten and more pronounced in the arms in the remaining two. In seven children, a positive Babinski's sign was the only sign of upper motor neuron affection.

### **Motor function**

Four of the 48 children were independent walkers, with a GMFCS level I-II, another six children were able to walk with assistive devices, i.e. level III, fourteen (21%) were classified at level IV and 27 (58%) at level V. For GMFM, the mean score for dimension A (lying and rolling) was 50%, for B (sitting) 30%, for C (crawling and kneeling) 20%, D (standing) 15% and E (walking, jumping and running) 11%. The mean total GMFM score was 26% (Figure 21 a). When it came to fine motor function, three children performed at BFMF level II, eight at level III, thirteen (27%) at level IV and 24 (50%) at level V.

### **Accompanying impairments**

Learning disability was present in 35 (73%). It was severe in 25 and mild in ten children. The percentage of children with learning disability increased with the severity of the motor disability ( $p < 0.001$ ). Six of the 38 children at GMFCS levels IV-V (16%) had no learning disability. Ten children were able to speak, but with dysarthria, while 38 (79%) with anarthria used augmentative and alternative communication such as Blissymbolics ( $n=11$ ), pictures, signing and/or body language to express themselves. All eleven children (23%) with a severe visual impairment were found among those performing at GMFCS level V. Epilepsy was present in 30 children (63%) (Figure 15).

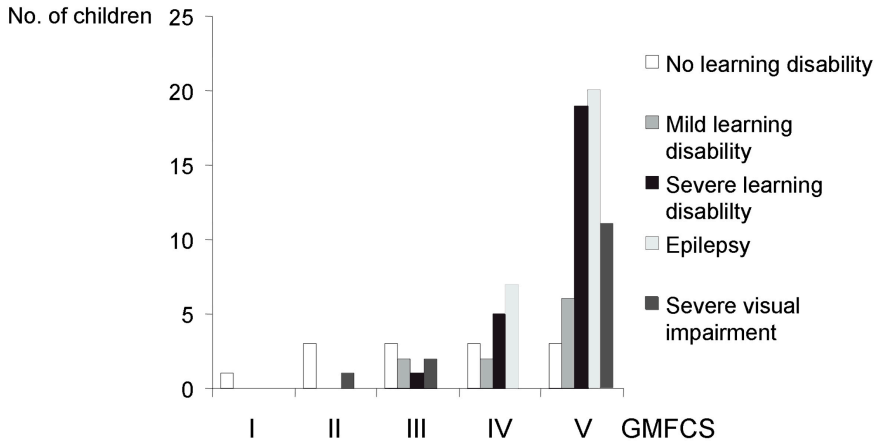


Figure 15. Learning disability, epilepsy and severe visual impairment by GMFCS level in 48 children with dyskinetic CP.

### Growth and nutrition

At birth, three of the 48 children (6%) had a body weight  $\leq -2$  SD for gestational age, but at follow-up the number of children with a body weight  $\leq -2$ SD had increased to 27 of 48 (56%;  $p < 0.001$ ). BMI was calculated in 47 children and was found to be less than  $-2$  SD in 14 (30%), all at GMFCS levels IV-V. None of the ten children with walking ability had a gastrostomy. The eighteen children who had a gastrostomy all performed at GMFCS level IV-V. Their mean weight deviation was  $-1.97$  SD, as compared to  $-3.4$  SD in the 20 children at these GMFCS levels without gastrostomy ( $p=0.011$ ; Figure 16). Nine of the 12 children with the most severe motor impairment who did not have a gastrostomy, had a BMI below  $-2$  SD, compared with none of the 13 children with a gastrostomy ( $p < 0.001$ ). Twenty of the 30 orally fed children were given an energy-enriched diet.

### Maternal and birth characteristics

A maternal disorder had been present in eight pregnancies; diabetes mellitus in three, fever at delivery in three, severe respiratory infection in one with hydrops fetalis in the near-term infant and prophylactic treatment against thrombosis with heparin in one. In the latter case, the child had an intracranial haemorrhage. Of the 48 investigated children, six (13%) were born before 34 completed weeks of gestation, four (8%) at 34-36 weeks and 38 (79%) were born at term. One child was SGA and six were LGA. Four children were twins. Five children were born at 42 completed weeks, four of whom were AGA (mean SD 0.68) and one LGA. For the 42 children born  $\geq 34$  weeks of gestation, peri/neonatal adverse events, with no history of prenatal compromise, had been present in 33 (81%). Placental

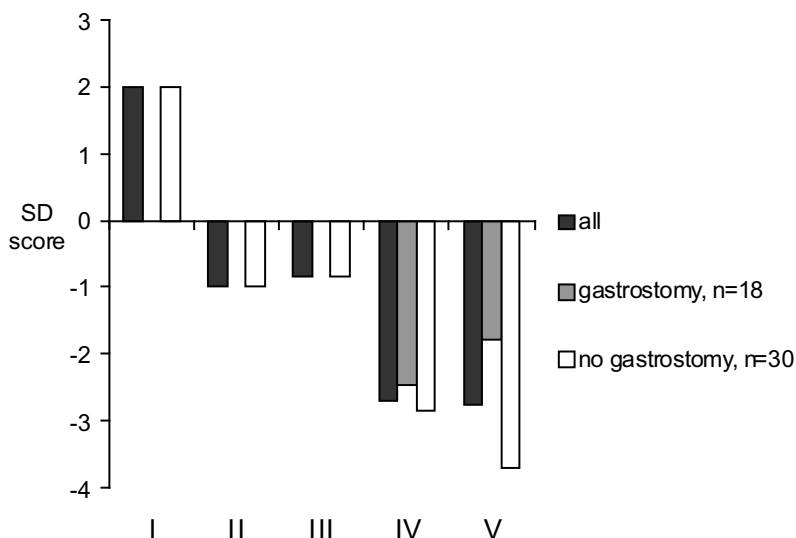


Figure 16. Weight deviation (SD) by GMFCS level at the time of investigation in 48 children with dyskinetic CP.

abruption and/or uterine rupture had occurred in eight (19%) compared with 1% in other CP types ( $p < 0.001$ ). Nineteen (45%) of the children required assisted ventilation compared with 12% with other CP types ( $p < 0.001$ ).

A selection of peri/neonatal factors adapted from Badawi (Badawi et al. 1998b) is listed in Table 21. When the strict criteria of ACOG were applied, 15 children met all the essential criteria (Table 22). The gross motor function in the 15 children who met all four essential criteria of ACOG was more severely impaired, with 13 of 15 (87%) performing at GMFCS levels IV-V, as compared to 19 of 27 (70%) in the other children with dyskinetic CP (ns) and 34 of 200 (17%) in those with other CP types born after  $\geq 34$  weeks of gestation ( $p < 0.001$ ; Figure 17).

### Neuroimaging

Of 39 children born after  $\geq 34$  weeks of gestation, pathological changes could be found in the basal ganglia and/or the thalamus in 30 (Table 23). In two of these children, the findings on CT or MRI could be referred to first and second-trimester lesions, i.e. pachygyria and basal ganglia calcification due to congenital cytomegalovirus infection in one and schizencephaly and haemorrhage in the basal ganglia in the other. In one child there was evidence of early third-trimester lesion, in 24 children there were late third-trimester lesions and in seven children there were both early and late third-trimester lesions.

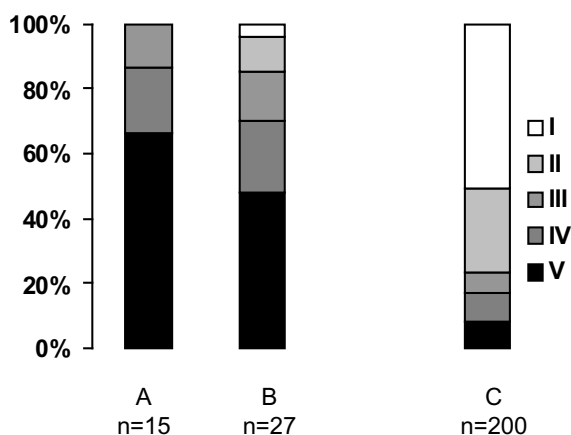


Figure 17. Distribution of gross motor function by GMFCS levels in children  $\geq 34$  weeks: Those meeting all four essential criteria of ACOG (A) remaining children of the dyskinetic group (B) and children with all other CP types (C) in the area born 1991-1998.

Table 21. Perinatal factors in term children and near term children with dyskinetic CP, born 1991-1998.

	Dyskinetic CP $\geq 34$ w n=42 (%)	Other CP types $\geq 34$ w n=200 (%)	All live born $\geq 34$ w n=199,051 (%)
Apgar score 5 min <5	28 (67)	15 (8)	591 (0.3)
Neonatal seizures	36 (83)	36 (18)	308 (0.2)
Spontaneous vaginal delivery	15 (36)	136 (68)	176,205 (89)
Instrumental vaginal delivery	8 (19)	14 (7)	11,086 (6)
Emergency caesarean section	19 (45)	36 (18)	10,549 (5)

### Changes in prevalence and severity of dyskinetic CP

The severity of motor impairment in dyskinetic CP varied over time. However, severe motor impairment has been most common during the study period. In the last three four-year cohorts, 75% or more of the group were confined to wheelchair ambulation. The prevalence and severity of motor impairment are shown in Figure 20 a.

Table 22. Essential criteria of The American College of Obstetricians and Gynecologists Task Force on Neonatal Encephalopathy and Cerebral Palsy for definition of an intrapartum event sufficient to cause cerebral palsy, applied on 42 children born  $\geq 34$  weeks with dyskinetic CP, by occurrence of peri/neonatal adverse event.

Children born $\geq 34$ w n=42	Children with peri/ neonatal adverse events n=33	Children with no peri/ neonatal adverse events n=9
No. of essential criteria		
4	15	0
3*	18	1
2	0	1
1	0	7

\*Sixteen children (15 of whom with peri/neonatal adverse event) failed to meet only the pH criterion, three of whom had pH  $\geq 7$  at delivery, while tests in eight children were performed after buffer treatment and more than 30 minutes after birth. In five children no analysis of pH was performed.

Table 23. Neuroimaging findings in 48 children with dyskinetic CP.

Stage of brain development	<34 w n=6	$\geq 34$ w n=42
1 <sup>st</sup> and 2 <sup>nd</sup> trimester		
Maldevelopment and basal ganglia lesion	0	2
3 <sup>rd</sup> trimester - early/mid		
White matter lesion only	2	1
- early and late		
Basal ganglia/thalamus/cortico-subcortical lesion combined with white matter lesion	1	7
- late		
Basal ganglia/thalamus lesion/cortico-subcortical lesion	1	24
Normal finding	0	5 (3 CT, 2 MRI)
Not done	2	3

#### IV. Bilateral spastic cerebral palsy – prevalence through four decades, motor function and growth

##### Birth years 1959-1998

After a significant decrease in the 1960s ( $p < 0.01$ ), the prevalence of bilateral spastic CP in the birth-year period 1967-1970 was 0.52 per 1,000 live born and rose to 1.27 in 1983-1986 ( $p < 0.01$ ), followed by another decrease to 0.69 per 1,000 live born ( $p < 0.01$ ) in children born in 1995-1998. The prevalence changes were significant for both the preterm and the full-term group (Figure 18). A contemporary steep decrease of perinatal mortality took place. The two preterm groups, i.e. those born at 32-36 weeks and those born before 32 weeks, followed a similar pattern but with a shift in time. The moderately preterm group, i.e. born after 32-36 weeks of gestation, peaked in the birth-year period 1979-1982, while the group born before 32 weeks peaked much later, in the birth-year period 1991-1994. In the term group after some increase in 1983-1990 the prevalence largely experienced a decreasing trend over the whole period ( $p < 0.01$ ). The percentage of children born at term decreased over the period ( $p < 0.05$ ) and after 1975 more children with bilateral spastic CP were born preterm than at full term. In the preterm group, the percentage of children born before 32 weeks of gestation rose from 14 of 36 (39%) in children born 1975-1978 to 29 of 41 (72%) born in 1995-1998 ( $p < 0.001$ ). In Figure 20 b the live birth prevalence and distribution of the severity of the gross motor impairment in the 763 children, 426 children (56%) of whom were born preterm, are presented.

After a rise from 1959 to 1986 in predominantly severe bilateral spastic CP ( $p < 0.001$ ), there was a significant decrease ( $p < 0.01$ ). The decrease of severity

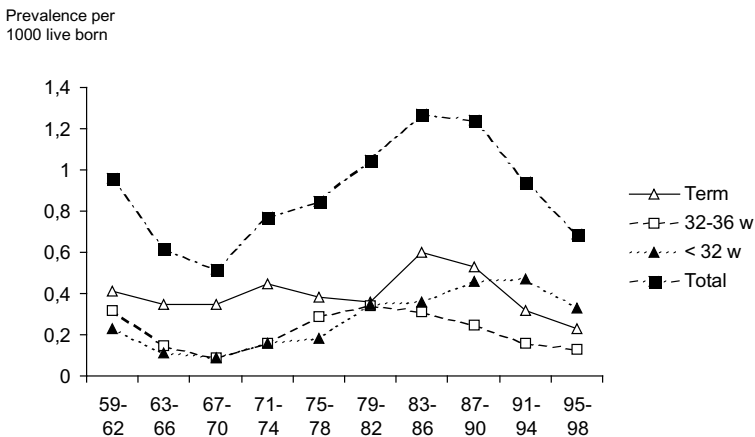


Figure 18. Crude prevalence of bilateral spastic CP in children born in 1959-1998. Of children with bilateral spastic CP, 337 children were born at term and 426 preterm, whereof 248 (58%) before 32 weeks of gestation.

mainly occurred in children born at term ( $p < 0.01$ ). No contemporary decrease in severity could be seen in preterm children. A larger percentage of the children born at term had a severe motor impairment than of those born preterm, 46% and 33% respectively ( $p < 0.001$ ). This difference persisted throughout the entire 40-year period. There was a male predominance in the preterm group with 59% boys ( $p < 0.01$ ). In both boys and girls there were larger percentages of severe motor impairment among those born at term (boys;  $p < 0.05$ , girls;  $p < 0.001$ ).

## Birth years 1991-1998

### Gross motor function

In the re-evaluation of the 167 children born in 1991-1998, the GMFCS level was modified in four cases, three of which had changed from GMFCS level III to II and one from III to IV. Fourteen percent performed at GMFCS level I, 34% at level II, 10% at level III, 25% at level IV and 17% at level V. Sixty-nine of 98 (70%) ambulatory children and 40 of 69 (58%) non-ambulatory children were born preterm. The distribution of GMFCS levels in the sub-groups of children in whom body weight, body height, GMFM and muscle tone data were analysed did not differ from the distribution in the whole group. The groups were therefore regarded as representative.

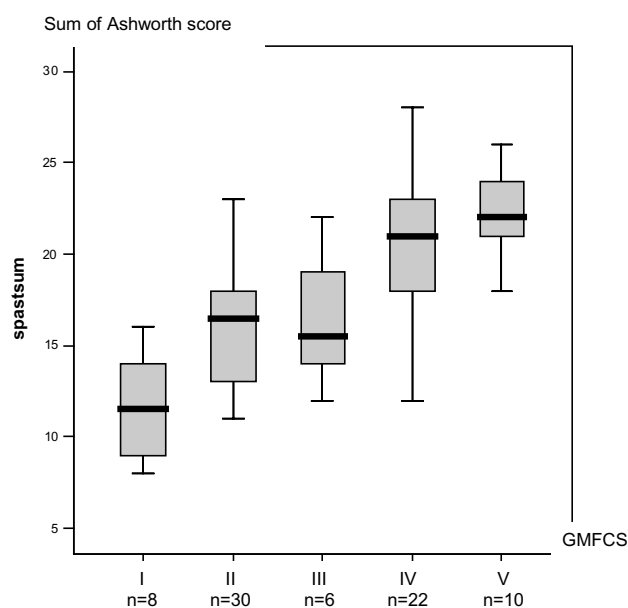


Figure 19. Median of spasticity score sum in eight muscle groups (adductors, hamstrings, quadriceps, and plantar flexors in both legs) by Gross Motor Function Classification System (GMFCS) level in 76 children. Boxes denote 25<sup>th</sup> to 75<sup>th</sup> percentile, whiskers extend from largest to smallest value. Maximum sum of Ashworth score 40.

The mean GMFM score at the age of four to 12 years (mean age 6.5 years) for the five dimensions and for the total score by GMFCS level in 100 children (60%) was analysed. There was a correlation between the GMFCS level and the total GMFM score, as well as between the GMFCS and the five separate GMFM dimensions (Spearman's rho 1.0;  $p < 0.01$ ).

### **Spasticity**

The Ashworth score, in eight selected muscle groups in the lower extremities was available in 76 (46%) of the 167 children (Figure 19). The distribution of a sum of Ashworth scores from eight muscles correlated with GMFCS ( $p < 0.01$ ). The median Ashworth score was 1 at GMFCS level I-II, 2 at GMFCS level III and 3 in GMFCS levels IV-V. The median Ashworth score for the knee extensors correlated with the GMFCS levels, (Spearman's rho 0.949;  $p = 0.014$ ).

### **Growth**

Information on body weight at four to 12 years of age (mean age 7.1 years) was available in 124 (74%) of the 167 children, representing 67% of the children at GMFCS level I, 72% at level II, 76% at level III, 76% at level IV and 82% of the children at level V. The weight deviation at follow-up correlated to the GMFCS level (Spearman's rho -0.975;  $p < 0.001$ ). The more severe the motor impairment, the more children weighed less than -2 SD ( $p < 0.02$ ). No such correlation was found between birth weight and GMFCS level. There was a significant difference between mean weight deviation at birth and at the time of follow-up, for preterm and term born children ( $p = 0.004$  and  $p = 0.005$ , respectively) (Table 24). There was a significant correlation between mean deviation of height at follow-up and GMFCS level in 106 children, (Spearman's rho -0.9,  $p = 0.037$ ), and a larger proportion of children with a body height  $< -2SD$  was found among those with a severe motor impairment, i.e. GMFCS IV-V ( $p < 0.05$ ). No correlation between severity of the motor impairment and BMI was found. Fifteen children had a BMI less than -2 SD, and six a BMI more than 2 SD (Table 25).

*Table 24. Mean weight deviation at birth and at the time of follow-up by GMFCS level in 76 children born preterm and 44 born at term.*

GMFCS	Preterm			Term		
	n	Mean birth weight deviation (SD)	Mean weight deviation at follow-up (SD)	n	Mean birth weight deviation (SD)	Mean weight deviation at follow-up (SD)
I	8	-0.56	-1.29	8	0.61	0.06
II	32	0.17	-0.77	8	-0.45	-1.08
III	6	0.08	-1.81	7	-0.41	-1.07
IV	20	0.21	-1.23	11	-0.15	-1.21
V	10	-0.35	-2.0	10	0.42	-0.95



Table 25. Mean height deviation and body mass index at the time of follow-up by Gross Motor Function Classification System level in 106 children with bilateral spastic CP.

GMFCS	n	Mean height deviation (SD)	Mean BMI deviation (SD)
I	14	-0.54	-0.13
II	37	-0.98	-0.41
III	9	-1.94	-0.56
IV	28	-1.46	-0.48
V	18	-1.68	-0.53

### Comparison of dyskinetic and bilateral spastic CP

The motor impairment outcome after comparable adverse events differed between dyskinetic and bilateral spastic CP (Table 26) and was more favourable in the near-term and term children with bilateral spastic CP than in those with dyskinetic CP ( $p < 0.01$ ).

Table 26. Outcome of severity of motor impairment in near term and term children with a history of perinatal adverse event, born in 1991-1998.

GMFCS	Bilateral spastic CP ≥ 34 w n=46	Dyskinetic CP ≥ 34 w n=39
I-III	31	11
IV-V	23	33

Risk factors such as low Apgar score, acidosis and neonatal seizures were more common in the dyskinetic group ( $p < 0.001$ ). In the near term and term children, 45% of the dyskinetic and 25% of the bilateral spastic children were born by emergency caesarean section ( $p < 0.05$ ; Table 27). The severity of the motor impairment varied over time in both CP types. The severely motor impaired group has dominated in dyskinetic CP (Figure 20 a and b). This subgroup has been the one varying in bilateral spastic CP, while the contribution of mild motor impairment has been more constant over time. Gross motor function, expressed as GMFM performance, followed the same pattern in the two CP types (Figure 21 a and b). Weight at follow-up showed a negative mean deviation for all GMFCS groups in both dyskinetic and bilateral spastic CP, with the exception of the only child performing at level I in the dyskinetic group (Figure 22 a and b).

Table 27. A comparison of occurrence of perinatal adverse events in children with bilateral spastic CP and dyskinetic CP born 1991-1998.

Perinatal event	Gestational age	Bilateral spastic CP n=167		Dyskinetic CP n=48	
		<34 w n=91 (%)	≥34 w n=76 (%)	<34 w n=6 (%)	≥34 w n=42 (%)
Apgar 5 min <5	≥34 w	12 (16)	28 (67)		
pH <7	≥34 w	2 (3)	17 (40)		
pH 7.0-7.1	≥34 w	5 (7)	3 (7)		
Abruptio placentae	<34 w	10 (11)	0 (0)		
	≥34 w	1 (1)	8 (19)		
Neonatal seizures	≥34 w	24 (32)	35 (83)		
Emergency caesarean section	<34 w	51 (56)	4 (67)		
	≥34 w	19 (25)	19 (45)		

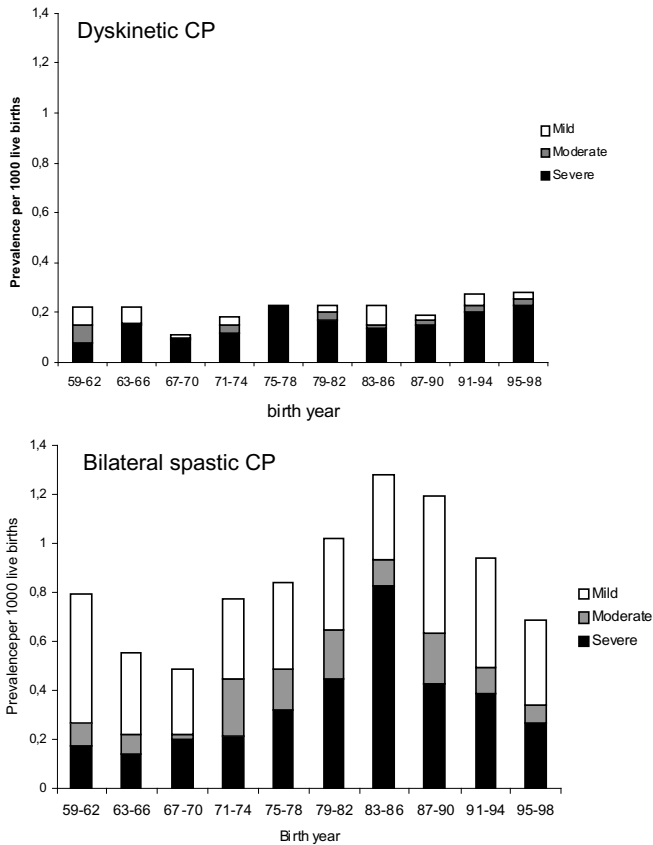


Figure 20 a and 20 b. Prevalence and severity of motor impairment in dyskinetic (a) and bilateral spastic CP (b) (mild: independent walking, moderate: walking with aids, severe: wheel-chair dependent)

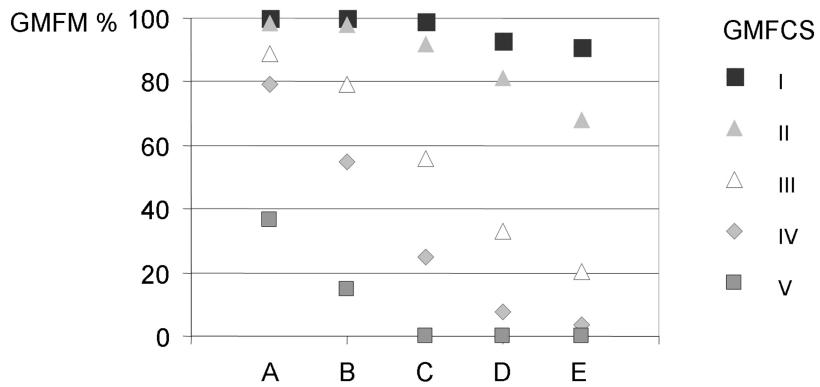
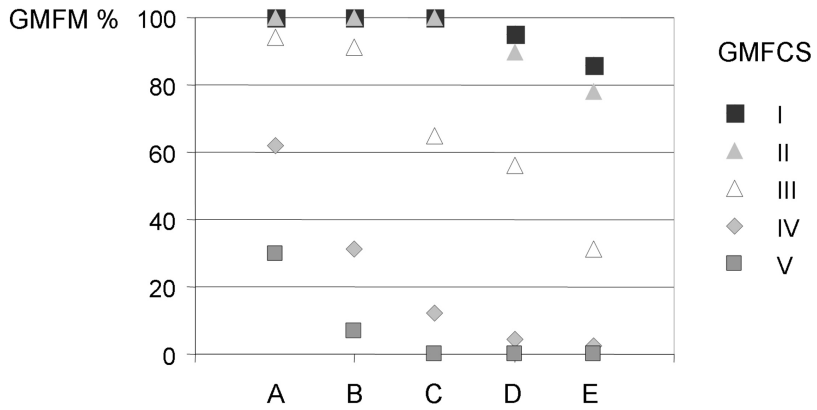


Figure 21 a and b. GMFM by GMFCS levels in 48 children with dyskinetic CP (a) and in 100 children with bilateral spastic CP (b). GMFM dimensions: A: lying and rolling, B: crawling and kneeling, C: sitting, D: standing, E: walking, running and jumping.

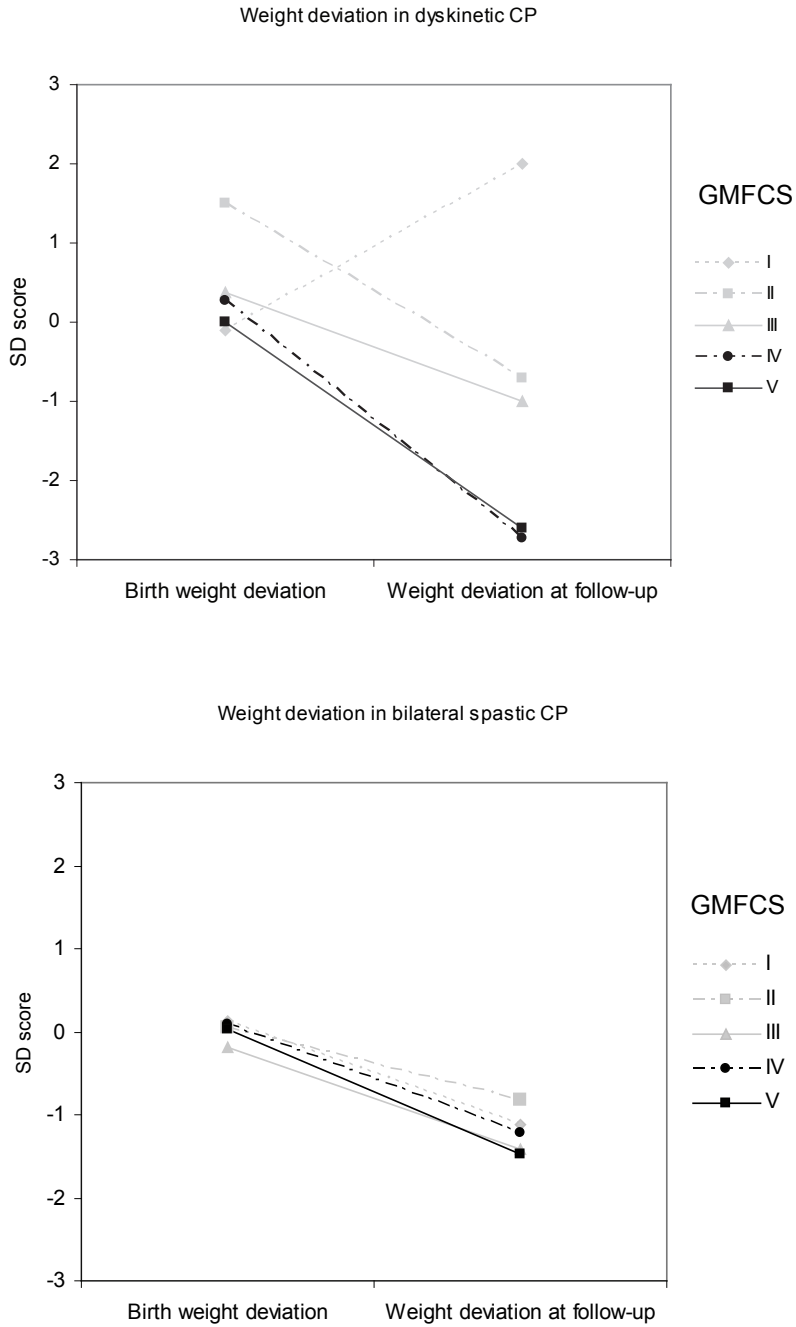


Figure 22 a and b. Weight deviation (SD) at birth and at follow-up at 5-13 years (mean 9) in dyskinetic CP and at 4-12 years (mean 7.1) in bilateral spastic CP.

## Discussion

The Panorama of Cerebral Palsy in Sweden Study comprises 1,770 subjects with CP from the four-decade birth-year period 1959 to 1998. During these years, perinatal mortality has decreased steeply and maternal and peri/neonatal care has undergone major changes. This has been accompanied by significant variations in CP prevalence, in this and other CP registers (Surman et al. 2003, Blair and Watson 2006). The decrease in CP prevalence since the mid-1980s in western Sweden has taken place in both the preterm and term groups. It was especially marked in very preterm children in the birth-year period 1995-1998, mainly in those with diplegic CP, which explains the higher relative frequency of hemiplegia among all cases of CP. However, diplegia was still the most frequent CP type in preterm children. The pronounced decrease in perinatal mortality coincided with an initial rise in predominantly preterm bilateral spastic CP, followed by a decrease in the same groups. Children born after 32-36 weeks were the first to follow this pattern, succeeded by those born before 32 weeks of gestation. Quite the opposite trend has been observed in Western Australia, where an increase in prevalence is seen in children born very and extremely preterm, although the overall trend is decreasing (Blair and Watson 2006).

Changes over time within the CP population are to be expected, as more preterm and more fragile children survive. At the same time the severity of impairment may be reduced with more advanced care. CP epidemiology is therefore important as an indicator of hazards, relevant to maternal, peri- and neonatal care. Major changes have in fact occurred over the years. Certain subgroups, such as cases due to rubella infection and rhesus immunisation, have more or less disappeared in the developed countries, while others, such as more cases with very low and extremely low gestational ages, have been added. Although the new preterm survivors constitute a high-risk population for various kinds of disability, including CP, the advances in neonatal intensive care have allowed far more infants to survive without CP than with CP (Hagberg et al. 1982). A comparison of perinatal mortality and crude prevalence of CP in the birth-year cohort 1983-1986, when the prevalence of CP reached its peak, and the latest cohort of 1995-1998, revealed that an additional 133 children would survive without CP, when translated into the number of live births in the latter period. This was despite the fact that the survival of preterm children had increased substantially. At variance with these results, in a similar comparison between the 1980s and 1990s in extremely preterm children, Wilson-Costello et al. reported a rise in neurodevelopmental disability contemporaneously with an increase in survival (Wilson-Costello et al. 2005). On the other hand, a decrease in both rate and severity in a corresponding population in the 1990s was reported by Surman (Surman et al. 2003), thereby supporting the findings in the present study.

The rise in the prevalence of dyskinetic CP from 0.17 per 1,000 live births in

the birth-year period 1983-1986 to 0.28 per 1,000 is a matter of concern. The vast majority of cases of dyskinetic CP were found among the children born at term. In contrast to the contemporary decrease in the overall CP prevalence in the area, the prevalence of dyskinetic CP was found to be at the same level as in the 1960s, despite developments in modern obstetric and neonatal care. The 55 children in the present study constituted 13% of all children with CP born in 1991 to 1998. This was a larger percentage than that reported from other centres in Europe (SCPE 2002). This finding was probably due to differences in classification. In many countries involved in the SCPE, the principle of diagnosing CP by the dominant syndrome has only recently been applied (SCPE 2002). As a result, children with dyskinetic CP and spasticity have more frequently been diagnosed as spastic CP. Had this been done in a similar way in this study, the percentage of dyskinetic CP would have been six per cent of all CP, a figure that would be more in accordance with the 6.5% in the SCPE report from 2002 (SCPE 2002). The children with dyskinetic CP were born at term in 79% and 2% SGA, in contrast to an earlier study from Sweden by Kyllerman, in which 65% were born at term and 10% were SGA (Kyllerman 1982).

We found a reduction over time in the percentage of children born at term in the bilateral spastic group and after 1975 preterm children dominated this group. The gross motor function of those born at term was poorer throughout the period. Although their motor impairment was more severe than that in children born preterm, the severity of the motor impairment decreased over the last 15-year period even in this group. This was in accordance with the findings reported from Western Australia by Blair and Watson (Blair and Watson 2006), where disability scores decreased but the more severe impairments were found among children born at term. Dolk et al. found similar trends in Northern Ireland (Dolk et al. 2006).

In this study, a positive trend was found between increasing gestational age and the percentage of all CP types with the mildest motor impairment. This was mostly due to the large group of children with hemiplegia or unilateral spastic CP. However, in this group, the trend in the 1990s was towards more severe gross and fine motor impairment. Although the children born at term dominate in the group with hemiplegia, the proportion of hemiplegia in the extremely preterm group had increased in the 1990s. It remains to be seen whether these trends persist.

Based on the criteria for aetiology used in this study, a prenatal aetiology was considered in 30% of the children born between 1995 and 1998, most of whom were born at term or near term. Forty per cent, constituting 35% of all children born at term and 49% of all preterm children were assigned to the peri/neonatal group. This high percentage of peri/neonatal aetiology has persisted over the last 12-year period, from 1987 to 1998. The morphology of brain injury is strongly related to the stages of brain development (Truwit et al. 1992, Barkovich et al. 2001, Flodmark et al. 2003, Krägeloh-Mann 2004). Several studies have shown that

insults at different stages of gestation are correlated to specific imaging findings (Truwit et al. 1992, Cowan et al. 2003) and that the lesions correspond to clinical pictures (Krägeloh-Mann et al. 2002, Krägeloh-Mann 2004). In the children born in 1995-1998, MRI and/or CT had been performed in no fewer than 85% of cases. The neuroimaging findings were similar to those in a recently published multi-centre study in Europe, where 72% could be affiliated to the third trimester as compared to 65% in western Sweden (Bax et al. 2006).

The most frequent finding at neuroimaging was periventricular atrophy, present in 40%, as compared to 43% in the study by Bax et al. (Bax et al. 2006). Maternal/foetal infection or inflammation as a cause or predisposing factor for the white-matter damage has been suggested in such cases (Grether et al. 2003, Jacobsson et al. 2002). In preterm children with a peri/neonatal aetiology, two-thirds were born very preterm or extremely preterm. In this group, the clinical criteria for the aetiological classification were intracerebral haemorrhage of grade III-IV in two-thirds of the cases.

All 31 children born at term in the peri/neonatal group had sustained HIE, intra-cerebral haemorrhage or CNS infection. There was an association between basal-ganglia and cortical/subcortical lesions, i.e. brain damage occurring late in the third trimester, and low Apgar scores in children born at term. This is in accordance with findings of Cowan et al., who studied 351 term infants with NE, seizures or both. More than 90% of those without specific syndromes or congenital defects had evidence of perinatally acquired insults. Damage to the basal ganglia, thalami and cortex was the most common pattern of severe acute perinatal injury (Cowan et al. 2003). In the present study, no fewer than 77% of the term children with peri/neonatal aetiology had suffered birth asphyxia considered severe enough to cause CP (MacLennan 1999). Moreover, in 58% the birth asphyxia was considered to have started intrapartum. In a Norwegian survey of subjects with CP born in 1970-1999, perinatal brain injury was considered to be the cause of CP in 39 of 99 children with a birth weight of > 2,500 g, 31 of whom had HIE (Meberg and Broch 2004). In a Canadian study, birth asphyxia was present in 22% (Shevell et al. 2003). Thornberg et al. reported moderate or severe HIE in 0.7 per 1,000 live-born term children (Thornberg et al. 1995) and Evans et al. in 1.61 per 1,000 births (Evans et al. 2001). Badawi et al. found that 13% of survivors of NE had CP, more severe and with more cognitive impairment and epilepsy than in children with CP without NE (Badawi et al. 2005). This was in accordance with our findings, i.e. that 68% of the children born at term or near term with dyskinetic CP had an Apgar score of < 5 at five minutes. This was significantly more common than the 20% recorded among all other children with CP born at term in 1991-1998. HIE had been present in 28% of near-term/term children with CP in these birth years. The association of asphyxia and HIE to CP has been opposed by many (Blair and Stanley 1988, Paneth 2001, Nelson 2003, Clark and Hanskins 2003). Medico-legal issues have been raised (Perlman 1997). The burden of proof rests heavily on the shoulders of those who propose such

an association. The strict criteria of ACOG (ACOG 2003) demand early investigation with acid-base status from arterial umbilical blood at delivery. Such an analysis may be overlooked in a critical resuscitation. This was the case in 11 of the dyskinetic children born  $\geq 34$  weeks. However, 15 met all the essential ACOG criteria and perinatal adverse event did not differ between the two groups. HIE in infants born term or near term was a major aetiological factor in dyskinetic CP in this study.

Over the years, there have been changes in the aetiological spectrum, gestational age distribution and the clinical picture of dyskinetic CP. In the study by Kyllerman (Kyllerman et al. 1982), hyperbilirubinemia was present as a contributory factor in 32%. Following the development of safe routines for testing and treatment, hyperbilirubinemia has almost disappeared as a risk factor for dyskinetic CP in children born in Sweden. The subgroup of children born preterm, who are more vulnerable to hyperbilirubinemia than those born at term, had decreased from 35% in the study by Kyllerman to 19% in the present one.

According to the criteria used in the present study, the aetiology was considered unclassifiable in 30% of cases, a somewhat larger percentage than in the previous birth-year period, 1991-94 (Hagberg et al. 2001). This may be due to a decrease in the incidence of associated post-haemorrhagic shunt-dependent hydrocephalus had occurred, indicating an improved outcome with fewer large intra-ventricular haemorrhages (Persson et al. 2005). The cases with less severe haemorrhages may more frequently have been assigned to the unclassifiable aetiology group, due to the strict criteria in this study. Intracerebral haemorrhage of lower severity, grade I-II, has been associated with neurodevelopmental impairment in children born preterm (Patra et al. 2006). More than half of the children in the unclassifiable group born in 1995-1998 had not been examined using MRI. Half of them had normal CT scans.

A further decrease in the percentage of children with CP of unknown aetiology may be expected when MRI becomes more widely used. However, in more than 10% there were normal results on neuroimaging in both the European multi-centre study (Bax et al. 2006) and in this study. In these cases, mechanisms that cannot be visualised with neuroimaging must be sought (Stanley et al. 2000).

In addition to the aetiological criteria which form the basis of this long-term study, the individual risk factors associated with CP may be considered. However, individual risk factors cannot be taken as a cause of CP, but rather as predisposing conditions making a foetus or newborn more susceptible to subsequent negative events (Stanley et al. 2000). Analyses of the presence of four conventional risk factors for CP were performed in this study. As in all studies of CP, low Apgar scores constituted a major risk factor (Stanley et al. 2000). Among children with CP born at term, Apgar scores of  $< 5$  at five minutes were 36 times more common than in the general population of children born at term in the area during the



same time period. A low Apgar score at five minutes among children born at term was strongly associated with HIE and hypoxic death (Hogan et al. 2006) and CP (Thorngren-Jerneck et al. 2001, Moster et al. 2001) has been reported, thereby, supporting the present findings. Whether a low Apgar score is a consequence of existing brain damage or a sign of a new insult leading to CP is debatable (Blair and Stanley 1988, Nelson and Grether 1999). In the present study, the latter supposition was supported by three independent observations. Firstly, the correlation found between low Apgar scores and dyskinetic CP, a syndrome which is known mainly to occur in children born at term or near term after severe asphyxia (Kyllerman et al. 1982, Krägeloh-Mann et al. 2002) and to be caused by lesions in the basal ganglia. Secondly, the finding of nearly normal or normal Apgar scores in children with CNS malformations dating from the first and second trimester and in children born at term with periventricular atrophy occurring late in the second or early in the third trimester. The children with a prenatal aetiology of CP did not signal any frailty in the form of depressed vital signs at or after birth. Thirdly, MRI findings were compatible with a late third-trimester, perinatal aetiology. This is in accordance with a report by Mercuri et al., who found basal ganglia and thalamus lesions in 72% of full-term children with a one-minute Apgar score of 0-3 and neonatal encephalopathy (Mercuri et al. 2002).

Maternal disorder included a variety of chronic or severe acute conditions during pregnancy. From the start of this CP study, it has been shown to be significantly associated with an increased risk of CP. In a case-control study of 149 children with hemiplegia born 1969-1978 and 445 matched controls, maternal disorder was found in 4.8% of term controls and 8.5% of controls born preterm (Uvebrant 1988). In the present study, a quarter of the mothers of children with CP had this risk factor, 17% of those in the term group and 35% of those in the preterm group, i.e. a four-fold increase in risk. The two most frequent conditions were fever at delivery and maternal diabetes. Antenatal factors such as chorioamnionitis have been associated with preterm labour and CP (Jacobsson and Hagberg 2004). In a case-control study comprising 148 children with spastic CP from birth cohorts in western Sweden born in 1983-1990 and 296 matched controls, infectious factors were found to be significantly yet weakly associated with CP (Jacobsson et al. 2002). Chorioamnionitis has also been proposed as a risk factor in near-term and term CP (Wu et al. 2003).

The percentage of multiple pregnancies has risen since the 1980s. This has been attributed to the enhanced opportunity for assisted fertilisation, increasing maternal age and higher survival rate at lower gestational ages (Stanley et al. 2000). In children born in 1995-1998, 14% of the children were multiples and more than 30% of them were born after assisted fertilisation (Hvidtjorn et al. 2006). The high percentage of multiple pregnancies may be expected to decrease as the policy of using fewer eggs in in-vitro fertilisation has become more widely accepted (Engmann et al. 2001). The antenatal death of co-twins is believed to increase the risk of CP in the surviving twin (Stanley et al. 2000); there were three such cases

in this study. From a survey of nine SCPE registers, Glinianaia et al. reported that an increased risk of CP in multiple birth was associated with growth deviation (Glinianaia et al. 2006). Topp et al., on the other hand, attributed the higher prevalence of CP in multiple births to preterm birth in another SCPE survey (Topp et al. 2004). The latter notion was opposed by the almost four-fold increased risk in term multiple birth compared to term singleton rate of CP in Western Australia (Blair and Watson 2006).

Deviation in intrauterine growth, SGA and recently also LGA are frequently discussed risk factors. An increased risk associated to SGA has been primarily demonstrated in term births (Kyllerman 1982, Blair and Stanley 1992, Uvebrant and Hagberg 1992) but in the present study of children born 1995-1998, only one child who was SGA was born at term. However, in a large population of CP cases, SGA and LGA were shown to be associated with an increased risk of CP in preterm births as well (Jarvis et al. 2003). Excluding multiple births from the children born 1995-1998, the incidence of SGA was 3.6%, which is somewhat but not significantly higher than the expected rate of 2.3%. Nor did LGA, present in 3.6%, differ statistically from the expected rate. If the above-mentioned risk factors had been applied, an additional 18 of the 45 (40%) unclassifiable cases could have been allocated to an aetiological group. The vast majority of these cases were considered to be prenatally derived.

Children born at term with peri/neonatal compromise, i.e. intracranial haemorrhage/stroke, cerebral infection and HIE, appeared to be particularly burdened when it came to the severity of the resulting fine and gross motor impairment, as well as the accompanying impairment load. Half of them developed spastic CP, while the other half developed dyskinetic CP. There was a significant difference in the severity of the motor impairment between the two groups; only 16% children with dyskinetic CP were able to walk, as compared to 67% of the children with spastic CP.

The classification of motor function by means of the GMFCS has improved the opportunity to describe gross motor function in a simple, comprehensive and uniform way in children with CP. It is important to note that this classification has made it possible to compare the panorama of CP within and between countries (SCPE 2000, 2002). It is recommended for use together with, and not as a replacement for, an agreed CP classification (Morris and Bartlett 2004). In the birth years 1991-1998, the distribution of gross motor function, expressed as the GMFCS, was similar to that reported by Nordmark et al., with 75% at GMFCS levels I to III and 25% at levels IV to V (Nordmark et al. 2001), corresponding to 70% and 31% in the present study. This is in accordance with a recent Australian study (Howard et al. 2005). The level of GMFCS was also found to correlate strongly to at least three accompanying impairments present in CP, i.e. learning disabilities, visual impairment and epilepsy. Consequently, the GMFCS can be used as an indicator of total disability load. The GMFCS classification remained

stable over time, in accordance with a recent report (Palisano et al. 2006). A change in GMFCS level affiliation was revealed in two percent of the children with bilateral spastic CP. The GMFCS can also serve as a framework for comparisons of measures of spasticity and gross motor function measures, one of which is the GMFM.

The GMFM is a valid measure that has acquired a widespread use since its introduction (Russell et al. 1989). However, one limitation is that the maximum achievement is that of a healthy five-year-old, which creates a ceiling effect in the results obtained by the most able children. For those with severe motor impairment, small achievements may not be possible to detect. In the children with dyskinetic and bilateral spastic CP, as in a study by Abel (Abel et al. 2003), performance in the five dimensions and the total score for the GMFM correlated with the level of GMFCS in comparable age spans.

When the new CP classification of bilateral spastic CP was applied, 21 of 23 children with tetraplegia were classified as bilateral spastic CP at GMFCS level V, while two were at GMFCS level IV but had a poorer hand function, at BFMF level V, thereby fulfilling the criterion for tetraplegia (Hagberg et al. 1975b). When it came to fine motor function, the distribution of BFMF in the different CP types was similar to that of the GMFCS. Although the overall pattern was similar, the correspondence between GMFCS and BFMF levels was no more than 57%, reflecting the different topographic profiles of the CP types.

Bilateral spastic CP and dyskinetic CP are large groups in the panorama of CP. They share the fact, that the motor impairment involves most of the body and the severity of motor impairment can range from GMFCS level I to V. Some children have both spastic and dyskinetic features, which may cause diagnostic difficulties. The accompanying impairment load is fairly similar when GMFCS level is taken into account. However, important differences, e.g. etiological period, gestational age at birth, the occurrence of perinatal adverse events and neurological findings, are apparent in the present study and in other studies (Krägeloh-Mann et al. 1993, 1994, 1995, Kyllerman et al. 1982, Kyllerman 1982). The signs of dyskinetic CP may be detectable at an early age (Einspieler et al. 2002) and are recognizable in gait analysis in those who walk (Davids et al. 1999). Dyskinetic CP was highly associated with lesions in the thalamus and basal ganglia in the present study. In accordance with this, Yokochi found involuntary athetoid movements in two-thirds of children with lesions of this kind (Yokochi 2004). Of the dyskinetic children in our study, 81% were confined to the dystonic group, corresponding to Kyllerman, who reported 70% (Kyllerman et al. 1982).

Spasticity is a characteristic in the majority of children with CP. It may be a dominant or additional sign. Although there still is a lack of consensus concerning its role in the motor function of CP, it is generally postulated that spasticity is related to the severity of the motor impairment (Sholtes et al. 2006). However,

the methods for measuring spasticity that are available to most clinicians, such as the Ashworth scale, are less accurate than instrumented measures used in clinical trials. Inconsistencies especially in the mid-range values have been reported (Damiano et al. 2002b). The finding in the present study of bilateral spastic children of a correlation between the degree of spasticity and the gross motor function, expressed as GMFCS level, supports that spasticity contributes to the motor impairment. However, motor function may also be impaired by weakness, lack of selective motor control (Giuliani 1991, Østensjø et al. 2004), balance problems (Rose et al. 2002), perception difficulties (Jacobson et al. 2002) and contractures (Hägglund et al. 2005a). Strength in particular has been shown to correlate well with motor function in walkers with spastic diplegia (Ross and Engsberg 2002) and to be possible to improve by training (Damiano et al. 1998, 2002a). In addition to physiotherapy (Barry 1996, Fowler et al. 2001, Bower et al. 2001, Dodd et al. 2002) including stretching (Pin et al. 2006), various treatments have been designed, e.g. selective dorsal rhizotomy (Peacock and Arens 1982), intrathecal baclofen (Albright 1991) and botulinum toxin (Cosgrove et al. 1994, Graham et al. 2000) in order to improve function and/or quality of life (Engsberg et al. 1998, 2006, Albright et al. 2003, Koman et al. 2001, Balkrishnan et al. 2004). Other interventions are orthopaedic surgery (Gage and Novacheck 2001) and oral pharmacotherapy (Verrotti et al. 2006). Observational reports are currently being replaced by controlled trials and long-term follow-up (Albright et al. 2003, Goldstein 2004, Hägglund et al. 2005a, Lannin et al. 2006, Engsberg et al. 2006). The benefit of long-term follow-up has been shown in southern Sweden, where the early detection of imminent hip luxation has successfully reduced the frequency of dislocated hips (Hägglund et al. 2005b).

The nature of accompanying impairments such as intellectual and neuropsychological function in CP is insufficiently known (Fennell and Dikel 2001). However, some evidence relating to the specific profiles in different CP types has emerged (Pueyo et al. 2003). The present study on gross and fine motor function and accompanying impairments focused on the four- to eight-year age band, an age at which a diagnosis of CP can be reliably certified and most paediatric accompanying impairments can be identified. Furthermore, these preschool and early school ages are important for planning and providing support for the optimal schooling of the child with CP. When accompanying impairments were considered, the children born before 28 weeks of gestation had the highest percentages of all impairments. This is in accordance with Marlow et al., who found that cognitive and neurological impairments were very common in children born before 26 weeks of gestation (Marlow et al. 2005). In a study by Lupton et al. almost 30% of infants with a mean gestational age of 26 weeks and normal head ultrasound had CP or learning disability at follow-up (Lupton et al. 2005).

Cognitive impairment in children born at  $\geq 34$  weeks was less common in children with dyskinetic CP than in bilateral spastic CP at the same GMFCS levels, even though it was frequent, especially in the dyskinetic non-walkers. Somewhat

larger percentages of learning disability and epilepsy were found than we have previously reported in the follow-up study. This could possibly be explained by the age at follow-up for dyskinetic CP, which was five to 13 years, compared with four to eight years in the previous reports (Hagberg et al. 2001, Beckung and Hagberg 2002), indicating an increase in impairment load by age. Obviously, epilepsy, as well as learning disability, may not be diagnosed at an early age. Low Apgar scores are associated with a higher frequency of epilepsy (Sun et al. 2006), which is in accordance with our findings. In a review of CP research, Koman et al. reported that more than 50% of children with CP in the eight- to 17-year age band can walk without aid, 25% cannot walk and 30% are cognitively impaired (Koman et al. 2004). The corresponding findings in the present study were 61% independent walkers, 31% not able to walk and 40% with learning disabilities. As Bax pointed out (Bax 2004), these differences may occur when wide or undefined age bands are used.

The concept of severe CP is created by the SCPE and has not previously been used in CP studies in western Sweden (SCPE 2002). It is defined as non-ambulant CP corresponding to GMFCS levels IV-V, with concomitant severe learning disability. In this study, the percentage of severe CP would have been 18%, compared with 20% in the SCPE survey (SCPE 2002). Half the children with dyskinetic CP and 26% of those with bilateral spastic CP in the present study fulfilled the criteria for severe CP according to the SCPE (SCPE 2000). However, a concept of this kind should be used with care, as some children with an isolated motor impairment may be extremely disabled. Consequently, the number of children with a severe disability may be underestimated if the SCPE definition of severe CP is used.

In contrast to children with primary speech and language disorders, the benefits of speech and language therapy has not been firmly established in children with CP (Law et al. 2003, Pennington et al. 2004). However, there are studies supporting this notion, albeit few and small (Pennington et al. 2004). In a longitudinal study of six severely impaired children with CP using augmentative and alternative communication (AAC), a positive development was observed for three years. However, the initial improvement was followed by a developmental arrest (Dahlgren Sandberg 2006). Difficulties in integrating AAC into school and home settings have been reported (Udwin and Yule 1990, 1991a, 1991b). In the present study of dyskinetic CP, the motor impairment was often more severe than the cognitive impairment, interfering with communication and other activities of daily life. Anarthria was present in about 80%, whereas the rest spoke with dysarthria. Poor speech intelligibility was a strong predictor of restrictive patterns of communication in families of children with CP (Pennington and McConachie 2001). Access to communication aids is crucial for these children. Otherwise, their true potential may be underestimated.

In the series of dyskinetic CP reported by Kyllerman, 10% of the children were

born SGA (Kyllerman 1982) as compared to only two per cent in the present study. The latter does not differ from the expected number in the population (Nicklasson et al. 1991). The postnatal growth, however, deviated significantly and 56% of the children with dyskinetic CP weighed below -2 SD at the age of five to 13 years. Those with gastrostomy had less weight deviation than those without gastrostomy. The same pattern was found in bilateral spastic CP, where a considerable number of the children were underweight at the time of follow-up at four to 12 years of age, in contrast to the distribution of birth weight. A correlation was also found between GMFCS levels and the deviation in body weight at the time of follow-up, but not with weight at birth, indicating that low body weight was an acquired problem. In this study, 28% of the children with bilateral spastic CP weighed below -2 SD and 31% had a height below -2 SD. In the dyskinetic children, an even more pronounced deviation was found. This is a high percentage and it is equal to the situation in developing countries such as Ethiopia or India, where more than 30% of children weigh below -2SD (de Onis et al. 2004). A corresponding pattern was not found for BMI. This may be explained by the fact that a low weight and a short stature in combination will result in a calculated BMI that is normal for age.

Measuring growth in children with CP is a challenge. Skin-fold measures are recommended by some (Samson-Fang and Stevenson 2000) but considered by others to overestimate the nutritional status, due to the muscle wasting in severely impaired children with CP. Muscle wasting takes place especially in the legs, representing a quarter of the body weight. A low body weight in non-ambulant children with spastic quadriplegia may therefore be explained by atrophy of leg muscles due to non-use (Kong and Wong 2005). Malnutrition due to feeding problems (Reilly et al. 1996) and sometimes increased energy demands (Hemingway et al. 2001, Johnston et al. 2004) are important factors for underweight in children with CP. Our findings were in accordance with a recent report on growth in 273 children with moderate to severe CP by Stevenson and co-workers (Stevenson et al. 2006). They stated that “bigger children with CP had better health and social participation than similar smaller children”. In severely impaired children with CP, a gastrostomy has proved to be an alternative feeding method (Sullivan et al. 2005, Stevenson et al. 2006). In addition to consequences for motor performance (Campanozzi et al. 2006) and other abilities (Samson-Fang et al. 2002), malnutrition is associated with poorer results in IQ tests in children without neurological disorders (Upudhyay et al. 1989, Agarwal et al. 1995, Walker et al. 2000). It is possible to speculate about whether the nutritional problems these children frequently experience contribute to their cognitive impairment.

The simultaneous significant decrease in neonatal mortality and CP prevalence between the birth-year periods 1991-1994 and 1995-1998 suggests that CP has been successfully prevented in a number of cases. In two recent reviews, however, the more or less constant total rate of CP in the USA was taken as an indicator that CP is not possible to prevent, given the current state of technology (Clark and

Hankins 2003, Nelson 2003). The authors argue that foetal monitoring and subsequent interventions such as Caesarean sections have had little, if any, impact on CP prevalence. There are improvements in technical surveillance of labour, but the potential effect on the prevalence of CP is not yet known (Norén et al. 2006). The potential for prevention must be further explored to limit the consequences for children experiencing a perinatal adverse event. Although the mechanisms of damage are well studied (Johnston and Hoon 2000, Johnston et al. 2001, Ferreira 2004, Vanucci and Hagberg 2004), the means to overcome them are scarce. The strategy of cooling the head or body has shown promising results in some studies of children born at term (Jacobs et al. 2003, Shankaran et al. 2005). In preterm births, Hagberg and Jacobsson considered glucocorticoids in single dose the most important prevention (Hagberg and Jacobsson 2005).

In the wait for future advances in prevention, the statement by Blair and Watson is valid: “The current longevity of the severely impaired suggests that as much effort and funding should be put into maximizing the quality of life of the severely impaired throughout their life span as is expended in ensuring that they have one” (Blair and Watson 2006). Prevention in a wider sense is important. This is true not only in terms of limiting the lesion, but also to limit the consequences of CP. The early recognition of malnutrition and sensory impairments and the stimulation of self-initiated communication, together with an up-to-date motor evaluation and well-timed interventions are prerequisites for participation. However, these concerns must not end during adolescence. Transition to adulthood means less service provision for all too many, even though reduced functional ability and secondary musculoskeletal problems, including pain and lack of social integration are common (Stevenson et al. 1997, Andersson and Mattsson 2001, Michelsen et al. 2006).





## Conclusions

In the changing panorama of CP in western Sweden during the period from 1995 to 1998, the decreasing trend from the previous period continued, both in children born at term and especially in those born preterm. However, the increase in dyskinetic CP in children born at term is a matter of concern. In this group, perinatal HIE had been present in the majority of the children.

The classification of CP should be based on CP type and motor function, as the two combine to produce a relevant indicator of the total impairment load. In addition, information about gestational age and peri/neonatal morbidity can provide aetiological and prognostic information. The concept of unilateral and bilateral spastic CP, combined with the GMFCS adds structure and comprehensibility to the CP classification.

Dyskinetic CP is a CP type dominated by term-born, AGA children who have frequently experienced peri/neonatal adverse events. These children often have severe disabilities. Underweight is common. As the lesions causing dyskinetic CP appear to occur in the perinatal period, further research is needed to address the potential for preventing this specific CP type.

The live-birth prevalence of bilateral spastic CP has decreased since the mid-1980s, parallel to a decrease in the severity of motor impairment. Since the mid-1970s, bilateral spastic CP has been dominated by children born preterm. The severity of the motor impairment, which correlates with the degree of spasticity, has decreased during the last 15 years. The percentage of children with underweight and stunting is substantial, in particular in those with a severe motor impairment.



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