THE PREVALENCE AND SURVIVAL OF CHILDREN WITH CEREBRAL PALSY IN BRITISH COLUMBIA

By

BENJAMIN CHAO-LIANG LAI
M.D., China Medical College, Taiwan, R.O.C.

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Department of Medicine

The University of British Columbia
Vancouver, Canada

Date April 30, 1999
ABSTRACT

The evidence that trends in the prevalence of cerebral palsy (CP) and survival rate of children with CP have increased in most industrialized countries over the past four decades was noted through systematic review of a number of current population-based studies. The biggest difficulty in evaluating these papers was the lack of a standardized protocol. Researchers working in different countries used different definitions, different criteria of inclusion and ascertainment, and different prognostic factors for survival. The information presented was not easily comparable and could not be readily extrapolated to the situation in British Columbia and Canada. As well, there was very limited information about the prevalence of CP and the survival of persons with CP and its associated problems in British Columbia.

To evaluate the prevalence of CP, survival rates, and associated problems in British Columbia, a retrospective study is proposed which links the information identified through two databases: the databases of the Health Surveillance Registry and the linked databases of the Centre for Health Services and Policy Research. Additional information relating to etiologic factors, functional abilities, prognostic factors and management will be extracted from the medical charts of patients using a data collection protocol designed as part of this thesis — the Cerebral Palsy Chart Review Protocol.

The feasibility of using the Cerebral Palsy Chart Review Protocol was tested on a convenience sample of hospital charts of children with CP. After reviewing sample charts, additional sections such as visual and hearing impairments were added, neonatal record evaluation forms were simplified, the sections on feeding and orthopaedic problems were modified, and the parameters for evaluating the severity of CP were expanded to include ambulation skills, mental ability, manual dexterity, and visual and hearing impairments.
Although the proposed provincial retrospective study has some limitations and methodological issues, overall the Chart Review Protocol is felt to be a feasible extraction method for a broader population review of cases. The proposed study will provide important information for health, education, social service and community planners and will be helpful in assessing methods and techniques in management and prevention of CP.
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ABBREVIATIONS

<table>
<thead>
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<th>Abbreviation</th>
<th>Description</th>
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<tbody>
<tr>
<td>BC</td>
<td>British Columbia</td>
</tr>
<tr>
<td>BCLHD</td>
<td>British Columbia Linked Health Database</td>
</tr>
<tr>
<td>CNS</td>
<td>central nervous system</td>
</tr>
<tr>
<td>CP</td>
<td>cerebral palsy</td>
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<tr>
<td>CT</td>
<td>computerized tomography</td>
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<tr>
<td>ELBW</td>
<td>extremely low birth weight</td>
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<tr>
<td>HIE</td>
<td>hypoxic-ischemic encephalopathy</td>
</tr>
<tr>
<td>HSR</td>
<td>Health Surveillance Registry</td>
</tr>
<tr>
<td>ICH</td>
<td>intracerebral hemorrhage</td>
</tr>
<tr>
<td>LBW</td>
<td>low birth weight</td>
</tr>
<tr>
<td>MLBW</td>
<td>moderately low birth weight</td>
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<tr>
<td>MRI</td>
<td>magnetic resonance imaging</td>
</tr>
<tr>
<td>MSP</td>
<td>medical services plan</td>
</tr>
<tr>
<td>PET</td>
<td>positron emission tomography</td>
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<tr>
<td>PNM</td>
<td>perinatal mortality</td>
</tr>
<tr>
<td>PVL</td>
<td>periventricular leukomalacia</td>
</tr>
<tr>
<td>SGA</td>
<td>small for gestational age</td>
</tr>
<tr>
<td>VLBW</td>
<td>very low birth weight</td>
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</table>
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Chapter 1. Introduction

Cerebral palsy (CP) is the term used to describe a movement disorder thought to be the result of non-progressive brain dysfunction caused by disordered development or by brain damage incurred during pregnancy, delivery, or in early life. Despite recent advances in medical care, CP is still the most common motor disorder of infants and children. More than 100,000 American children and adolescents are estimated to have neurological disabilities linked to CP and the conditions associated with it, particularly mental retardation (Newacheck and Taylor, 1992). Approximately 25 percent of people with CP identified by registries in France and the United Kingdom are unable to walk (even with assistance), and 30 percent are classified as mentally retarded (Evans et al., 1990; Rumeau-Rouquette et al., 1992); whereas, about 20 percent of persons with CP in Canada are moderately to severely mental retarded (Crichton et al., 1995). In addition to the high economic cost of caring for people with CP, emotional suffering and lost opportunities add immeasurably to the burdens of affected families (Kokkonen et al., 1991; Cox and Lambrenos, 1992; Hallum and Krumboltz, 1993).

Although CP has been described for more than one hundred years, the pathophysiologic mechanisms that underlie most of the CP syndromes remain poorly understood. Because the injury is central in origin, the motor involvement is commonly accompanied by other central nervous system (CNS) impairments, which may be cognitive, sensory, or communicative. However, because motor function is the earliest assessable developmental process, CP is usually the first identifiable developmental disability. The child suspected of having CP requires not only an investigation for etiology but also a comprehensive evaluation to detect impairment in other CNS functions. This enables directed interdisciplinary therapy with the goal of improving the child’s abilities and quality of life.
During the past three decades, improvements in obstetrical and neonatal care have dramatically reduced perinatal mortality among children of low birth weight (<2500g), very low birth weight (<1500g), and extremely low birth weight (<1000 g) (Hagberg et al., 1993, 1996; O'Shea et al., 1997, 1998; Pharoah et al., 1990, 1996; Topp et al., 1997). This has resulted in an increasing prevalence of CP and changing patterns of CP as well as influencing the survival rate of children with CP (Evans et al., 1990; Hutton et al., 1994; Crichton et al., 1995). Along with the improved neonatal care, there also has been marked improvement in the care of children with CP over the past 30 years. The children have access to improved health care, and new technologies allow them to have a better quality of life and fewer complications. Meanwhile, social support systems have assisted parents in caring for these children in their home or with the assistance of associate or foster families. As a result, most of these children no longer live in institutions but live in their communities and have a higher survival rate. This should prompt a reappraisal of their long term needs as well as their medical care plans. At present, however, there is very limited information about the prevalence of CP and its associated problems in British Columbia. While some preliminary work has been done regarding the survival of persons with CP, this work is not sufficiently detailed to provide the information required.

The objectives of this thesis are as follows:

1. To review various aspects of CP such as historical background, definition, epidemiology, etiology, classification, associated conditions, pathological findings, treatment, prevention, and outcome.

2. To evaluate the changing trends in prevalence, survival and associated variables of CP through systematic review of population-based journal articles located through several computerized bibliographic databases from 1966 to 1998.
3. To design a data extraction form, the Cerebral Palsy Chart Review Protocol, and to assess the feasibility of this protocol by conducting a pilot study.

4. To utilize epidemiological methods to develop a proposal for establishing the prevalence and survival of children with CP in BC.
Chapter 2. General review of cerebral palsy

To proceed with a series of proposed studies regarding the prevalence and survival of CP and its associated problems, it is essential to have a detailed understanding of CP. This chapter provides an overall review of CP.

2.1. Historical background

Motor disability in childhood has been recognized since the earliest of recorded history. However, CP was not distinguished from other motor disabilities until the 19th century when William John Little, an orthopaedic surgeon, attributed spastic rigidity to obstetrical complications and prenatal anoxia (Accardo, 1989). He described a specific type of CP known as spastic diplegia, which is still referred to as Little’s disease. In Little’s classic report of 200 cases, in 1861, he suggested specific etiologies for spastic diplegia and thereby offered an opportunity for preventive measures to be taken.

Since Little’s pioneering work further classification of various CP syndromes has occurred. William Osler introduced the term “cerebral palsy”, a contraction of the word “paralysis” derived from the German — Cerebral Kinderlahmung (cerebral child paralysis), in his 1889 book, “The Cerebral Palsies of Children”. He described the clinical findings in 150 cases of CP, grouped them according to presumed etiology, and speculated upon the pathophysiological mechanisms of CP. In 1897, Sigmund Freud (Accardo, 1982; Freud 1968), a child and adult neurologist before turning to psychoanalysis, introduced an early clinical classification in his classic text, Infantile Cerebral Palsy, which provided a basis for later classification and emphasized the existence of associated problems such as mental retardation, epilepsy, and visual disturbance. In regard to etiology, Freud emphasized prenatal influences in
suggesting that CP might be linked to "symptoms of deeper lying influence which have dominated the development of the fetus."

After World War II, advances in genetic and metabolic studies permitted delineation of disorders previously categorized under the broad heading of "cerebral palsy". For example, a specific etiology, hyperbilirubinemia, was established for a choreoathetoid CP syndrome. In 1956, the current agreed upon classification of the clinical motor disorders was adopted by the American Academy of Cerebral Palsy (Minear, 1956). Later, refinements in classification were introduced and summarized by Crothers and Paine (1959). Also, in recent decades, epidemiological studies were launched in the industrial developed countries. Efforts at prevention and early treatment of children with CP were initiated at the beginning of the twentieth century when programs in physical therapy were introduced. Extensive efforts have been focused on therapy, both for movement disorder and associated problems, as well as for psychological adaptation by the child and family.

2.2. Definition

CP is a symptom complex, rather than a specific disease. In 1957, a conference was assembled to address issues of terminology and classification of CP. From these discussions, the following definition was proposed:

"Cerebral palsy is a persistent but not unchanging disorder of movement and posture, appearing in the early years of life and due to a nonprogressive disorder of the brain, the result of interference during its development" (MacKeith, 1959).

Later, in 1964, a group of professionals met to review the terminology and classification of CP. The group reached agreement on a definition of CP as: "A disorder of movement and posture due to a defect or lesion of the immature brain" (Bax, 1964). The most recent consensus
definition states that: “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 its development” (Mutch et al., 1992). Children with CP caused by postneonatal events (after 28 days of age), such as meningoencephalitis or head trauma, are usually considered as having acquired or secondary CP, compared with those born with idiopathic or primary CP. The cerebral lesion of CP is static and nonprogressive, but changes do occur as the central nervous system matures, and the peripheral physical symptoms may change with brain development. For example, a hypotonic infant becomes a spastic or rigid child, and a child who is originally diagnosed as choreoathetoid may subsequently become dystonic and develop contractures. Consequently, precise definitions of CP and sufficient follow-up periods (at least 4 to 5 years) are important for identifying the type of CP exhibited and are a requirement for any accurate epidemiological study of CP.

2.3. Epidemiology

The prevalence of moderately severe or severe CP is estimated to be 0.9 to 3.0 per 1000 live births (Hagberg et al., 1993, 1996; Pharoah et al., 1990, 1996; Topp et al., 1997). When milder cases are considered, prevalence may be as high as 1.0 to 6.0 per 1000 live births. Estimates based on data from registers of children eligible for services (Evans et al., 1990; Grether et al., 1992) tend to be lower than those obtained from prospective cohort studies (Stanley and Watson, 1992; Pharoah et al., 1990) because, among other reasons, registers are likely to miss mild cases that can be diagnosed only by neurologic examination.

During the past three decades, improvements in obstetrical and neonatal care have highly increased the survival of very low birthweight infants. As very low birthweight infants are at high risk for CP, the rate of subsequent CP is 25 to 31 times higher among infants with a birth
weight less than 1500 grams when contrasted to full-sized newborns. Meanwhile, those whose birth weight is less than 2500 grams make up about 35 to 55% of all infants who show signs of CP later (Pharoah et al., 1990, 1996; Hagberg et al., 1993, 1996).

The trends in the prevalence of CP have also changed along with the decreasing perinatal mortality rate in the past three decades. The crude prevalence of CP declined from the 1950s through the 1960s, and then increased progressively in the 1970s and 1980s in most developed industrial countries, except in Norway and Australia (Meberg et al., 1990, 1995; Stanley and Watson, 1988, 1992). The prevalence of low birthweight CP increased in most current studies; and the lower the birthweight group, the later the rise in the prevalence of CP occurred (Pharoah et al., 1996). More detailed information about the changing trends in the prevalence of CP and associated disorders will be discussed in the next chapter.

2.4. Etiology

In 1861, William John Little postulated that the motor defects resulted directly from difficulties in the birth process. This opinion was held for over one century. Among some early critics, Sigmund Freud speculated that perinatal difficulties were the result of pre-existing abnormalities in the fetus rather than the cause of CP. The question of which came first, the brain damage or the asphyxia, was eventually resolved by the National Collaborative Perinatal Project (NCPP). This was a prospective study with a 7 year follow-up in order to determine the incidence and risk factors contributing to the development of CP. Of the children enrolled in the study, 0.5% were diagnosed as having CP by 3 years of age. Many of the infants who sustained asphyxia had congenital brain anomalies, while asphyxia alone accounted for less than 10% of all subjects with CP (Blair and Stanley, 1985; Nelson and Ellenberg, 1986). This supported Freud’s view that CNS abnormalities preceded birth in most children with CP. Meanwhile,
analyses of the NCPP (Nelson and Ellenberg, 1986; Nelson, 1988) found that the majority of persons with CP did not have specifically defined causes. Like etiologically unidentified mental retardation syndromes and congenital malformations, a specific etiology was lacking.

Like most disorders, CP has multiple risk factors, both causes and modifiers. It is associated with a variety of risk factors occurring before pregnancy, during pregnancy, or during the perinatal period (Kuban and Leviton, 1994; Torfs et al., 1990). Factors occurring before pregnancy include long intervals between menses and a history of spontaneous abortion and stillbirth (ibid). Factors occurring during pregnancy include congenital malformation, fetal growth retardation, twin gestation, and abnormal fetal presentation. During labor and delivery, there is an association with premature separation of the placenta and nonvertex or face presentations. Moreover, a recent study reported that decreased gestation and meconium-stained amniotic fluid were the only antenatal factors associated with increased odds for both death and CP (Spinillo et al., 1997). The abnormal presentation may be a marker for pre-existing difficulties in developmental maturation rather than being a cause of CP. During the early postnatal period, newborn encephalopathy may be linked to CP.

Many diseases and conditions can injure the developing brain and lead to CP; however, approximately one quarter to one third of all cases still have no definable cause. Problems during intrauterine development account for the largest proportion of known causes of CP (Naeye et al., 1989; Paneth, 1984, 1986). There are some common antecedents of CP, such as prematurity, asphyxia, prenatal abnormalities, biochemical abnormalities, genetic causes, environmental toxins, congenital infections, and postnatal events, but it is important to emphasize that most children with these known risk factors do not develop this disability (Eicher and Bastshaw, 1993). In Blair and Stanley’s study (1993), half (51.4%) of the children with spastic CP did not exhibit any identified risk factors.
It is clear that CP is not a single entity and the etiology of its various presentations is varied. The most common etiology is periventricular leukomalacia associated with prematurity (Kuban and Leviton, 1994). Yet in early childhood, infections of the CNS, stroke, head injury, or poisoning may be etiological factors. CNS infectious disorders in the first year have been reported to account for 6% of cases (Naeye et al., 1989). About 2% of persons with CP have a genetic etiology (Hughes and Newton, 1992). Although early studies had placed primary emphasis on perinatal asphyxia as causative, later studies (Nelson, 1988; Blair, 1988) found asphyxia neonatorum in only 3% to 13% of cases; even in quadriplegias only 14% of cases were due to birth asphyxia. In Powell’s study (1988a, 1988b), intrapartum events were most often associated with hemiplegia; whereas in diplegia, which is most often associated with prematurity, factors involved in the rate of fetal development were more important.

Some studies attribute brain damage in neonatal encephalopathy to the excess production of excitatory amino acids during the first postnatal days (Vannucci, 1990; Ford, 1990; Espinoza and Parer, 1991; Riikonen et al., 1992). In addition, disorders of neuronal migration may account for movement disorders and may lead to increased vulnerability to other kinds of injury. Immature neurones migrate to the cortical plate between 7 and 16 weeks of gestational age. Abnormal migration may be associated with reduction in cell size, and early neuronal death may occur. Such abnormalities may subsequently affect the development of association pathways and lead to motor dysfunction, seizure, and learning and behavioral problems. Environmental factors at sensitive developmental periods and genetic factors may affect neuronal migration (Harris, 1995).
2.5. Classification

As a diagnostic entity, "CP" makes no inference as to etiology, pathophysiology, degree of handicap, therapy, or prognosis. Despite multiple classification systems for CP, no one system has proved to be entirely successful. The multiaxial classification used to classify CP includes the type of dysfunction (physiologic), the location of the dysfunction (topographic), and associated conditions (supplemental) (Bax, 1964; Minear, 1956). Etiological classifications are unsatisfactory as are neuropathological classifications because similar clinical syndromes appear to result from different etiologies and are associated with different neuropathological findings. Since the abnormalities of muscle tone in early infancy may change over several years the initial classification must be tentative until the clear presentation of the syndrome can be identified. The classification may be difficult because of subtle abnormalities and mixed features that may be demonstrated on the motor examination.

The classification currently in use is modified from a system designed by the American Academy for Cerebral Palsy, as recommended by Minear (1956). The physiological classification is of value as a marker for associated deficits, including spastic (pyramidal), dyskinetic (nonspastic, or extrapyramidal), rigid, ataxic, tremor, atonic, mixed, and unclassified CP. The topographic classification is used only for the spastic types.

Differences among spastics are usually based on the number of limbs involved (topography) rather than on the quality of the movement disorder. Primitive reflexes may persist, leading to problems in posture and movement. In monoplegia, a relatively rare form of spasticity usually considered to be a variation of hemiplegia, only one limb of the body is affected. In hemiplegia one lateral half of the body is affected and the arm is usually more affected than the leg. In paraplegia only the legs are involved, with normal upper limb function. In diplegia all four limbs are involved, but the upper limbs may have only minimal involvement.
Triplegia, a three limb involvement, is often considered as a variation of quadriplegia and like monoplegia is quite rare. In quadriplegia all limbs are affected, and there is generally severe dysfunction of the four extremities. Infants with these various types are often described as floppy because of their initial hypotonicity. With increasing age, they show increased muscle tone and associated musculoskeletal deformities. The degree of mental retardation is usually higher proportionate to the number of limbs involved.

Two subgroups of extrapyramidal CP have been identified, dystonic and dyskinetic (choreoathetoid). An increase in the abnormal involuntary movements is associated with activity, with emotions, and when active muscle tension is necessary, but during sleep or relaxation, symptoms may be less intense. Primitive reflexes are more apparent in these forms. Dyskinetic cases with athetosis have involuntary movements that involve various muscle groups. As a result, they may appear contorted, stiff, or in continuous motion. Their speech may be dysarthric, and they are more likely to have feeding problems. Athetoid patients are usually associated with chorea, and are known as choreoathetoid. Their movements are relatively rapid and irregular and are more pronounced during voluntary movement. The dystonic form involves extremes of athetoid movement in which the body and/or extremities are forced into fixed postures by strong muscle contraction. The cases of dyskinetic CP are thought to have damage to the basal ganglion and to the cranial nerves; the cerebrum is usually less involved. Therefore, in these cases, there is less cognitive impairment than in those with spastic type.

When both extrapyramidal and pyramidal forms present in the same person, this is described as mixed type. The other types, ataxic, rigid, and atonic, are much less common. Patients with ataxias have a disturbance of the coordination of voluntary movements due to muscle dyssynergia. These patients may be hypotonic during the first 2 or 3 years of life.
2.6. Associated disabilities

In addition to problems with movement and posture, almost all children with CP have at least one additional disability associated with damage of the CNS (Jones, 1975). The most common of these associated deficits are mental retardation, sensory deficits, communication disorders, seizures, feeding problems, and behavioral or emotional problems. In general, the more limbs involved, the greater the degree of spasticity and the higher the likelihood of associated deficits.

Some reports found that certain associated features accompany particular types of CP (Capute and Accardo, 1991; Grether et al., 1992). For example, spastic hemiplegia is more often associated with seizures, hemianopsia, growth arrest, and cortical sensory deficits including visual abnormalities. Strabismus is manifested by 43% of spastic diplegia patients and seizure is present in 27% of the children with spastic diplegia (Ingram, 1995). Spastic quadriplegia is associated with epilepsy, mental retardation, dysarthria, and strabismus. Quadriplegia is more likely to be associated with seizures, extrapyramidal abnormalities, and severe cognitive impairment than hemiplegia or diplegia. Choreaathetosis is associated with mental retardation, auditory impairment, and dysarthria.

The most disabling problems for children with CP are the associated cognitive difficulties. Approximately one third to two thirds of children with CP have mental retardation (Evans et al., 1990; Rumeau-Rouquette et al., 1992). Among those with normal intelligence, most have perceptual impairments that place them at risk for learning disabilities. Children with hemiplegia have the best intellectual outcome, with more than 60% having normal intelligence. In contrast, less than 30% of children with spastic quadriplegia or mixed-type CP have normal intelligence.
Sensory deficits include both vision and hearing problems. Visual deficits, which occur in about 45 to 65% of children with CP, include refractive errors, field defects, amblyopia, nystagmus, and abnormalities of visual pursuit. Strabismus is found in more than half of all children with CP (Schenk-Rootlieb et al., 1992). Hearing loss occurs in about 10% of children with CP, predominantly in those whose disability derives from prenatal or postnatal CNS infections (Cohen et al., 1988). Associated learning disability is often a consequence of deficits in visual perception, in auditory processing, and in phonological sequence. Similarly, communication disorders in children with CP include speech motor deficits and central processing problems. Speech motor problems are common in extrapyramidal CP with dysarthria.

Approximately one quarter to one third of children with CP develop a seizure disorder (Aksu, 1990; Crichton et al., 1995). Seizures have been noted to be most frequent in spastic hemiplegia (55% to 72% of cases) and least common in choreoathetosis and the ataxic form (23%). Most seizures manifest by 2 years of age. As in the general population, tonic-clonic and partial complex seizure predominate.

2.7. Pathological finding

2.7.1. Full-term infants

Cerebral injury in the distribution of the middle cerebral artery is the most common finding in pathologically confirmed patients with hemiplegic spastic CP (Ment et al., 1984; Levy et al., 1985) and in patients with hemiplegia evaluated by computerized tomography (CT) and magnetic resonance imaging (MRI) (Wiklund et al., 1991; van Bogaert et al., 1992). For reasons that are not yet clear, isolated right-sided hemiplegia occurs twice as frequently as isolated left-side hemiplegia (Grether et al., 1992). Some children with hemiplegic CP have
periventricular atrophy, suggesting the presence of abnormalities in the white matter, and about one sixth have gross malformations of cerebral development (Wiklund et al., 1991). The CT or MRI scan is normal in another one quarter to one third of children with presumed congenital hemiplegia. The lack of recognizable areas of injury or abnormality provides support for the notion that some CP cases are related to abnormalities of brain development at the microscopic level, and diminishes the likelihood that the disease is caused by injury to a normally developed brain. Associated with the quadriplegic form are cavities that communicate with the lateral ventricles, multiple cystic lesions in the white matter, diffuse cortical atrophy, and hydrocephalus (Truwit et al., 1992).

In choreoathetoid and dystonic forms of CP, the microscopical appearance of the basal ganglia often resembles marble (status marmoratus) (Burn and Kyllerman, 1979). Persistently hypotonic, or atonic, CP implies the involvement of cerebellar pathways. An enlarged ventricular system is the most frequent finding of neuroimaging.

2.7.2. Infants born prematurely

During the past 30 years, neuropathologists have suggested that the periventricular white matter may be the site of the most important abnormality leading to congenital motor dysfunction. Periventricular leukomalacia (PVL) is the name given to the lesions characterized by foci of coagulative necrosis in the white matter near lateral ventricles (Banker and Larroche, 1962; DeReuck et al., 1982), and it is known as the predominant cause of spastic diplegia among moderately preterm infants with CP (Kuban and Leviton, 1994). Prominent hypoechoic periventricular areas on ultrasonographic imaging studies predict the later development of motor dysfunction (Leviton and Paneth, 1990). The MRI images in a preterm infants with CP are characterized by prolonged T2 signals (especially in the periventricular areas), distortion of
the normal contours of the lateral ventricle, and ventriculomegaly (Krageloh-Mann et al., 1995; Sugimoto et al., 1995; Rogers et al., 1994). Each of these MRI characteristics is probably a consequence of PVL. Premature infants who have ventriculomegaly without macrocephaly, even if they have never had ultrasonographically documented hyperechoic or hypoechoic periventricular abnormalities, also appear to have an increased risk of CP (Saliba et al., 1990; Weisglas-Kuperus et al., 1992).

A recent study (Olsen et al., 1997) regarding the finding of PVL by MRI and its clinical correlation in children found that the prevalence of PVL among all children born prematurely was 32%. PVL was observed in all children with CP, in 25% with minor neurological dysfunction, and in 25% of the clinically healthy preterm children. None of the children born at term had evidence of PVL.

2.8. Diagnosis

The diagnosis of CP rests on a clear understanding of the defined dimensions of the condition. There are three components to this definition. First, CP is a disorder of movement and posture. Second, CP is a result of abnormalities in the early development of the brain. Third, the disorder of CP must be nonprogressive. CP is a developmental disability and the diagnosis of a developmental disability is based on an analysis of the behavioral and the functional characteristics of a particular child. This is accomplished by partitioning the developmental processes into several functional domains (Pellegrino and Dormans, 1998). These functional domains fall into two major groupings: basic physiological and psychological processes and integrated functional processes. The first grouping includes sensory function, cognitive processes, and motor function. The second grouping is based on the first and includes communication/socialization skills, daily living skills, and mobility skills. CP is defined simply in
terms of decrements in motor function and mobility skills. Deficits in the other processes are defined as associated impairments and disabilities.

The pediatric assessment of the motor-delayed child begins with confirmation and quantification of “delay” by comparison of the infant’s performance with known motor milestones. This is best achieved through the use of historical information and observation.

There are three components for the motor examination of infants with delayed attainment of gross motor skills: (1) traditional neurological examination, (2) assessment of primitive reflexes, and (3) elicitation of postural response. Each component does not reliably detect CP except in extreme cases, but combining the parts of the motor examination aids the detection of CP.

Signs of CP can be detected by traditional neurological examination. However, the traditional neurological examination is of limited use for the early detection of CP because of instability of signs, changing interpretations of similar signs with age, and poor localizing value. In contrast to the adult and older child, the neurological examination of the infant changes with maturation. For instance, spastic diplegias may be initially hypotonic in the legs, whereas children with choreoathetoid CP may show generalized hypotonia before exhibiting their movement disorders. Furthermore, some children with CP may lose function, especially during the early second decade of life. This usually relates to the development of contractures, excess weight gain, or lack of motivation. Similar findings have different significance when they occur at different ages.

Primitive reflexes are brainstem-mediated reflexes that are present at birth and that diminish in activity during the first year of life (Capute et al., 1984). These responses have been associated with the development of normal motor activity (Capute et al., 1982) and are considered to be the earliest indicators of significant motor disabilities. Except for the most
extreme cases, isolated primitive reflexes do not portend motor handicap. Additional research is needed to determine the clinical applicability of combinations of reflexes.

Postural responses include righting and equilibrium responses. Postural responses become more evident while primitive reflexes diminish in activity. The volitional motor activity is closely associated with the appearance of postural responses.

Laboratory tests are not necessary to confirm the diagnosis. However, they can be helpful in establishing an etiology and suggesting a prognosis. Advances in the use of non-invasive brain-imaging techniques are now being utilized to improve assessment methods. Cranial ultrasound through the open anterior fontanelle is used in the early detection of PVL and other structural anomalies in infants. CT and MRI may also be utilized to evaluate cerebral structural changes (Volpe, 1992). In the future, assessments of metabolic activities and blood flows, by techniques such as using positron emission tomography (PET), may become more generally available (Chugani, 1992).

It is not sufficient to perform only a motor examination on the child who presents with “motor delay”. For example, mental retardation, communication disorders, and learning disabilities may present as motor delay. Psychological testing should be done to assess intellectual functions and a comprehensive neurodevelopmental examination, including assessment of language, cognition, and adaptive skills, in addition to motor skills, is indicated for the child who manifests gross motor delay.

Generally, according to the definition of CP, a child must have an obvious motor deficit for the diagnosis of CP to be considered. The chief complaint usually is that the child is not reaching motor milestones at the normal time. A careful history must establish that the child is not losing function, ensuring that the patient does not have a progressive disease. This history,
combined with a neurological examination establishing that the patient's motor deficit is caused by a cerebral abnormality, leads to the diagnosis of CP.

2.9. Treatment

After a complete developmental diagnosis, which includes investigation of associated dysfunctions in addition to the motor disability, the treatment of a child with CP is best accomplished by a team of knowledgeable individuals with different expertise. Before initiating treatment of a child with CP, quantifiable short-term and long-term objectives should be defined. Many factors must be considered in setting goals for children with CP, including CP factors, child factors, family factors, and environmental factors.

The goal of therapy is generally twofold: (1) to maintain functions, and (2) to maximize existing functions or develop new functions. Activities directed toward maintenance of function may include motor therapies to prevent contractures, positioning to ensure symmetry and prevent scoliosis, and soft tissue surgery to prevent hip dislocation. Activities directed toward maximization of functions and for the development of new skills focus on communicating effectively, on ambulatory and nonambulatory mobility, and on performing activities of daily independent living.

Nonmotor activities are frequently directed toward maintenance of self-esteem and prevention of secondary behavioral disturbances. Such activities include parental counselling to establish appropriate expectations, school placement consonant with the child's abilities, and opportunities for social interaction.

In most patients the diagnosis of CP is established during the first 2 years of life. Early intervention programs, which provide not only specific hands-on therapies but also psychological support, are thought to be beneficial, although there is no evidence that they
enhance the child’s development (Binder and Eng, 1989; Palmer et al., 1990). The concept of early intervention programs has even been questioned in a study of parent satisfaction with an infant stimulation program for CP. Russman (1998) also reported that early intervention programs to enhance motor and cognitive development in the physically handicapped population have not been shown to be beneficial, in contrast to early intervention programs for the environmentally deprived population.

It is important to develop a treatment plan together with family members. If they do not agree with a plan, or are financially or emotionally unable to follow through with it, the entire program will fail. If possible, the therapeutic program should be carried out in the home of the infants as part of an early intervention program (Palmer et al., 1990).

Therapies may include the use of medication for seizure, spasticity (Armstrong, 1992; Cosgrove et al., 1994), constipation, or gastroesophageal reflux. Neurosurgeons also may become involved, especially in certain cases of spastic diplegia in which the use of selective dorsal rhizotomy has been advocated to decrease spasticity (Dormans, 1993; Hendricks-Ferguson and Ortman, 1995). Even once an effective treatment program has been initiated, there is the need for periodic reassessment.

2.10. Prevention

During the past decades, numerous advances have been made in the prevention of CP. Immunizations, both active vaccination for rubella and rubeola and passive administration with anti-RH immunoglobulin to prevent Rh sensitization and the subsequent hemolytic disease of the newborn with kernicterus, have clearly altered the incidence and types of CP. Advanced techniques and improved care in the area of obstetrics and neonatology have provided the meaningful ability to manage both mothers and infants. For example, ultrasound permits
visualization of the fetus and placenta, measurement of certain physiological factors, and the establishment of the duration of pregnancy; amniocentesis permits detection of chromosomal abnormalities, culture of fetal cells to detect certain metabolic derangements, and detection of neural-tube defects through maternal alpha-fetoprotein analysis.

Low birthweight infants account for the greatest number of patients with CP. In preterm newborns, PVL has been associated with physiologic instability in the form of low blood pressure, ventilatory problems, and infection (Leviton and Paneth, 1990). Meanwhile, Spinillo and colleagues (1997) reported that decreasing gestation and the presence of meconium stained amniotic fluid were the only antenatal factors associated with increased odds for both death and CP in preterm infants. Thus, it would appear that further improvements in neonatal care may partly reduce the risk of PVL and consequently of CP in preterm infants.

Surfactant was introduced and routinely utilized for very preterm infants starting in the late 1980s. Some studies report that the decline in mortality among very low birth weight infants in the late 1980s and early 1990s has not resulted in an increase in the prevalence of CP (O’Shea et al., 1997, 1998; Hamvas et al., 1996; Palta et al., 1994; Schwartz et al., 1994). The increased neonatal survival and the decreased risk of CP, after introduction of surfactant, implies that brain damage attributable to complications of prematurity has decreased.

O’Shea and colleagues (1992) found a decreased risk of subarachnoid hemorrhage and intraventricular hemorrhage in babies whose mothers had multiple gestations, were preeclamptic, received tocolytic agents, and received steroids. On the other hand, Kuban and colleagues (1992) noted that the incidence of CP in infants whose mothers who were toxemic and had received magnesium sulfate was less than a comparable group. Meanwhile, Nelson and Grether (1995) reported that only 7.1% of mothers receiving magnesium sulfate gave birth to
babies who developed CP as opposed to 30% who did not receive the drug. These observations need confirmation with carefully controlled clinical trials.

2.11. Outcome

The data from some recent studies (Kudrjavcev et al., 1985; Evans et al., 1990; Hutton et al., 1994; Crichton et al., 1995; Strauss et al., 1998) show that the factors influencing survival of CP include immobility, severe mental retardation, manual dexterity, epilepsy, birth weight, and gestational age. There are also some factors which might influence the length of survival and quality of life, such as orthopaedic, visual and hearing impairments, respiratory infection, feeding problems, and technical advances and improvements in the management of these problems.

Although the life expectancy of severely involved individuals is significantly less than that of the general population, more than 90% of children with CP live to adulthood (Evans et al., 1990). The outlook varies slightly with the topography of the CP. In general, the more limbs involved, the worse the prognosis. In Hutton’s study (1994), the survival rate of mildly and even moderately affected children approached that of normal, unaffected children, at least for the first 20 years. Even severely disabled children had about a 50% probability of surviving to 20 years of age. Moreover, in Crichton’s study (1995), the overall survival rate at 30 years is at least 87 percent. Survival of persons with CP and associated problems will be discussed at length in Chapter 4.

2.12. Summary

Since Little distinguished CP from other motor disabilities in 1861, numerous definitions for CP have been proposed. The most recent consensus definition of CP states that “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 its
development” (Mutch et al., 1992). Etiological studies suggest that CNS abnormalities precede
birth for most children with CP, but CP is a syndrome, not a single entity, and the etiology of its
various presentations is varied. Because different etiologies may result in similar clinical
syndromes classification of CP remains difficult. Currently, CP is multiaxially classified
according to type, location, and associated conditions of the dysfunction.

Over the last few decades numerous advances have been made in the prevention of CP.
These include immunization to prevent Rh sensitization, administration of magnesium sulfate to
preeclampsia pregnant mothers and routine utilization of surfactant for extremely preterm or
low birth weight infants. Treatment of CP is generally limited to therapy designed to maintain
and maximize existing functions. Survival studies suggest that while numerous factors affect
survival and quality of life, the overall survival rate is high. Since over 100000 Americans are
estimated to have neurological disabilities linked to CP and since the condition has a high social
cost, the need for continued study and research cannot be overemphasized.

3.1. Introduction

The prevalence of cerebral palsy (CP) has generally increased in the past three decades (Riikonen et al., 1989; Pharoah et al., 1996; Hagberg et al., 1993; Topp et al., 1997). Most of this increase is attributable to a marked improvement in the survival of very small infants, brought about by improved perinatal fetal monitoring and improved neonatal intensive care involving routine use of mechanical ventilation and total parenteral nutrition (Cummins et al., 1993; Kuban and Leviton, 1994; Stanley, 1992). Since premature and low birth weight babies account for 35 to 55% of all CP cases, the increased prevalence rate of CP reflects increased survival as a result of advances in perinatal and neonatal care (Takeshita et al., 1989; Pharoah et al., 1996).

Some recent studies (Meberg, 1990; Meberg and Broch, 1995; Kavcic and Perat, 1998) appear to show a reversal of the increase of incidence in CP trends, although Stanley and Watson (1988,1992) reported that there were no definite changes in the prevalence of CP in Western Australia. Along with changing trends in the crude prevalence of CP, some associated issues of CP, such as the proportion of CP by birthweight, trends in etiological groups, trends in clinical type of CP, and trends in severity of CP, also appear to have changed. However, researchers working in different countries use different definitions, different criteria of inclusion, and different methods for ascertainment. Meanwhile, there is not enough information in these papers to fully explain the apparent change in trends.

The changing trends in the prevalence of CP naturally have important implications for health, education and social services. The purpose of this chapter is to examine the trends in CP
and to utilize epidemiological methods to evaluate population-based studies of the prevalence of CP and associated issues.

3.2. Data Sources and Methods

For this review study, only the time-trend reports or serial reports in which the data of the prevalence of CP were collected from the same region were included. The articles were searched for from several computerized bibliographic databases including MEDLINE, EMBASE, CINAHIL, HealthSTAR, Biologic Abstract, and Current Contents in OVID Database. Two groups of search terms were used. One was CP and prevalence, CP and incidence, and CP and epidemiology; the other was CP and preterm, CP and prematurity, and CP and low birth weight. Furthermore, to explore the etiologies and types of CP, additional groups of search terms used included CP and etiology or cause, and CP and classification or type.

Regional population-based articles published in English were selected from these databases from 1966 to 1998. Editorials, commentaries, letters, unpublished studies, abstracts, reports appearing only in government publications, and summaries of presentations given at medical meetings or brief transcripts of meetings were excluded (Escobar et al., 1991). Only data provided in the reports were considered; and no attempt was made to contact the authors for missing data.

The review was limited to articles describing population-based registries of CP in which attempts were made to ascertain from multiple sources all cases with CP in a region. Hospital-based studies were not included unless all births in a region occurred at the hospital. Studies which did not calculate the prevalence of CP by using the total number of livebirths in
the region as the denominator were excluded. Studies in which the population was based on specified groups (e.g., very low birthweight infants, multiple births) were also excluded.

The scientific quality of the selected articles was assessed according to criteria of applicability and validity developed by Oxman and Guyatt (1988), Oxman et al. (1991), and Mulrow (1987). The elements for evaluating applicability were modified to focus on specific purpose, definitions, criteria of inclusion and exclusion, ascertainment of case, sample size, methods of statistical analysis, and summary or conclusion. Validity was evaluated for elements such as role of chance, biases, confounding. In addition, although they did not meet all our criteria, several recent papers which raised important points or issues related to the prevalence of CP were also reviewed.

3.3. Results

Searching through several computerized bibliographic databases from 1966 to 1998, a number of studies relating to prevalence or incidence of CP were noted. However, only a small number of articles or serial reports were compatible with our criteria: i.e. population-based time-trend studies on the prevalence of CP (Dowding and Barry, 1988; Hagberg et al., 1975, 1979, 1984, 1989, 1993, 1996; Kavic and Perat, 1998; Meberg and Broch, 1990, 1995; Pharoah et al., 1987, 1990, 1996; Riikonen et al., 1989; Stanley and Watson, 1979, 1988, 1992; Takeshita et al., 1989; Topp et al., 1997). The number of citations and details of ineligible studies identified through different databases are presented in Table 3.1. There was overlap between different databases; therefore, the number of cases unique to any one given database is given in brackets in Table 3.1. Although MacGillivray and Campbell’s report relating to the changing pattern of CP in Avon met our criteria, it is not included in this study because the denominator population was calculated with all births of the region (MacGillivray and Campbell,
1995). It did not mention the proportion of stillbirths, and consequently the results of the prevalence of CP would be lower than those whose denominator population was calculated with all livebirths.

These studies were mostly performed in Northern and Western Europe. The characteristics of each study are shown in Table 3.2. The studies lasted from 6 to 32 years, and were all done some time between 1954 and 1990. All livebirths born to a mother residing in the study region were utilized as the denominator population for the prevalence of CP in all studies. Infants weighing less than 500g were excluded in Meberg’s study (1990, 1995) because of the likelihood for low validity of these data.

Subjects in most studies were ascertained from multiple medical data sources (Table 3.2); however, in Riikonen’s study (1989) all cases with suspected CP in the study region were examined by a pediatrician or a child neurologist at a regional university central hospital. The diagnosis of each case was confirmed by at least 3 years of follow up, mostly by 5 years. Most cases who died with a definite diagnosis of CP before the end of the follow up period were included; however, deaths before 2 years of age were excluded in Hagberg’s study (1975, 1979, 1984, 1989, 1993, 1996) and deaths before one year of age were excluded in Topp’s study (1997). In Kavcic and Perat’s study (1998), children with CP who migrated into or out of Slovonia between 1981 and 1995 were not included, neither were those who had died. As the prevalence of CP was usually evaluated with the subjects who were born in the study area, the exclusion of emigrant cases lowered the prevalence. Postneonatal cases were excluded in all except two studies. In Hagberg’s study, postnatal cases damaged before two years of age were included, and in Riikonen’s study (1989), postnatal cases up to 3 years of age were also included.
Some studies contained a relatively small number of cases and therefore were sensitive to chance variation. However, they achieved high validity of data because of stability of the population, centralized programs for follow-up, and methods used for collecting the data which would have secured nearly complete ascertainment. Alternatively, some serial studies showed the strength of their project in consistency of methodology with regard to investigators, search methods, clinical classification and criteria for inclusion. Possible errors can therefore be expected to be systematic and evenly distributed over time, and should thus not influence the general trends.

**Proportion of CP by gender**


**Proportion of CP by birthweight**

The proportion of infants with CP weighing less than 2500g at birth in each study is shown in Table 3.3. The proportion of low birthweight infants for all individuals with CP increased from 10-35% in the 1960s to 35-55% in the 1980s. For instance, the changing distribution of low birthweight infants with CP among the total number of children with CP in the Tottori cohort (Takeshita et al., 1989) dramatically increased during the past 30 years, from about 10% before 1960 to over 50% after 1980. Similarly, the most recent years in Pharoah's study showed that over 50% of all infants with CP were of low birthweight and that those with a birthweight of less than 1000g recently made an important contribution to the total; whereas in the earlier years of their study, low birthweight cases constituted only 25-35% of the total
Table 3.1. Number of citations and details of ineligible studies identified through different databases.

<table>
<thead>
<tr>
<th></th>
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<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Total number of citations</td>
<td>130(130)</td>
<td>61(23)</td>
<td>76(5)</td>
<td>40(21)</td>
<td>22(8)</td>
<td>187</td>
</tr>
<tr>
<td>Total eligible studies</td>
<td>18(18)</td>
<td>12(1)</td>
<td>10(0)</td>
<td>3(0)</td>
<td>5(0)</td>
<td>19</td>
</tr>
<tr>
<td>Total ineligible studies</td>
<td>112(112)</td>
<td>49(22)</td>
<td>66(5)</td>
<td>37(21)</td>
<td>17(8)</td>
<td>168</td>
</tr>
<tr>
<td>1) Not a regional population-based study, or the prevalence of CP was not calculated by using the total number of livebirths in the region as the denominator.</td>
<td>8(8)</td>
<td>4(2)</td>
<td>5(0)</td>
<td>10(6)</td>
<td>6(1)</td>
<td>17</td>
</tr>
<tr>
<td>2) Not time-trend or serial reports of the prevalence of CP.</td>
<td>23(23)</td>
<td>5(1)</td>
<td>11(1)</td>
<td>5(3)</td>
<td>4(1)</td>
<td>29</td>
</tr>
<tr>
<td>3) The population of the study was based on specified groups.</td>
<td>39(39)</td>
<td>18(11)</td>
<td>26(3)</td>
<td>22(14)</td>
<td>12(6)</td>
<td>73</td>
</tr>
<tr>
<td>4) The studies were focused on postneonatal or acquired CP or on adults with CP.</td>
<td>2(2)</td>
<td>2(1)</td>
<td>1(0)</td>
<td>0(0)</td>
<td>0(0)</td>
<td>3</td>
</tr>
<tr>
<td>5) Not written in English.</td>
<td>28(28)</td>
<td>9(3)</td>
<td>17(0)</td>
<td>8(5)</td>
<td>1(1)</td>
<td>37</td>
</tr>
<tr>
<td>6) Reports were editorials, letters, commentaries, unpublished studies, abstracts, and reviews, appearing only in government publications, summaries of presentations at medical meetings or brief transcripts of meetings.</td>
<td>27(27)</td>
<td>15(6)</td>
<td>14(1)</td>
<td>3(0)</td>
<td>2(1)</td>
<td>35</td>
</tr>
</tbody>
</table>
Table 3.1. (Continue)

**Note:** 1. Obviously some studies show up in more than one database. The numbers in brackets indicate studies not found in the previously listed databases. Since MEDILINE is first listed, the number in brackets for MEDILINE is always identical to the total number of cases found.
2. The number in the "total" column is the total number of different studies identified through all databases.
3. 38 of the EMBASE citations are the same as the MEDLINE.
4. 69 of the HealthStar citations are the same as the MEDLINE.
5. 18 of the Biological Abstracts citations are the same as the MEDLINE.
6. 12 of the Current Contents citations are the same as the MEDLINE.
7. 25 of the HealthStar citations are the same as the EMBASE.
8. 12 of the Biological Abstracts citations are the same as the EMBASE.
9. 9 of the Current Contents citations are the same as the EMBASE.
10. 16 of the Biological Abstracts citations are the same as the HealthStar.
11. 10 of the Current Contents citations are the same as the HealthStar.
12. 8 of the Current Contents citations are the same as the Biological Abstracts.
13. 19 citations were ineligible by two criteria.
14. 4 citations were ineligible by three criteria.
15. No eligible citations were found using the CINAHL, the Health and Psychosocial Instruments, and the Cochrane Database of Systematic Reviews.
Table 3.2. Characteristics of studies in the prevalence of cerebral palsy (CP).

<table>
<thead>
<tr>
<th>Authors (year)</th>
<th>Region and period of study</th>
<th>Denominator population</th>
<th>Ascertainment of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dowding and Barry (1988)</td>
<td>Eastern Health Board area of Ireland 1976 - 1981</td>
<td>All resident births in three counties of the Eastern Health Board of Ireland. Total population 1.19 million in 1981 with over 25000 births per year.</td>
<td>Multiple ascertainment for resident birth children with CP at 4 years of age, and those who died with a definite diagnosis of CP before this age. Postneonatal cases were excluded.</td>
</tr>
<tr>
<td>Hagberg et al. (1975, 1979, 1984, 1989, 1993, 1996)</td>
<td>Southwest region of Sweden 1954 - 1990</td>
<td>All livebirths in the southwest region of Sweden with a total population of 1.7 million inhabitants, and livebirths during the birth year period 1987 - 96 were 91542.</td>
<td>At least 4-year-old cases were identified from multiple sources including residents at the time of the study. Deaths before 2 years of age were excluded. Postnatal cases removed before two years of age were included.</td>
</tr>
<tr>
<td>Kavcic and Perat (1998)</td>
<td>Slovenia 1981-1990</td>
<td>All livebirths in the study area between 1981 and 1990, and who still resided there in 1995. Total population is about 2 million with 22000 to 28000 annual livebirths.</td>
<td>The cases were ascertained by physicians from multiple sources. Mild forms of CP were identified up to age 5 years. Postneonatal cases were excluded. Children with CP who migrated into or out of the area during 1995 or before were not included, neither were those who had died.</td>
</tr>
<tr>
<td>Meberg (1990) and Broch (1995)</td>
<td>Vestfold County, Norway 1970 - 1989</td>
<td>All livebirths ≥500g in the county of Vestfold, Norway. Total births weighing 500g or more through 1970 to 1989 were 45976.</td>
<td>Resident birth cases were traced through active research from multiple sources. At least 4 years follow up before a definitive diagnosis. Postneonatal cases were excluded.</td>
</tr>
<tr>
<td>Pharoah et al. (1987, 1990, 1996)</td>
<td>Mersey region of UK 1966 - 1989</td>
<td>All livebirths in Mersey region of UK, according to the area of maternal residence at the time of birth. Annual livebirths were around 30000 to 45000 during 1966 to 1984.</td>
<td>Several sources of information were used: handicap registers, family claiming disability allowance, schools for disabled, and all death certificates. Postneonatal cases were excluded. Case ascertainment was completed until 5 years of age.</td>
</tr>
<tr>
<td>Riikonen et al. (1989)</td>
<td>Turku, Finland 1968 - 1982</td>
<td>All livebirths in Turku region. Total population is about 450000, approximate 5500 deliveries a year.</td>
<td>All cases with suspected CP were examined by physician at the regional hospital. All cases were followed up for at least 5 years. Postnatal cases until 3 years of age were included.</td>
</tr>
<tr>
<td>Stanley and Watson (1979, 1988, 1992)</td>
<td>Western Australia 1956 - 1985</td>
<td>All livebirths in Western Australia identified by birth registrations (1967 - 74), and midwives' notifications (1975 - 85).</td>
<td>Cases were ascertainment from multiple sources and updated to the age of 5 years. Postneonatal cases were excluded. It is not reported whether deaths were included if CP was known to be present.</td>
</tr>
<tr>
<td>Takeshita et al. (1989)</td>
<td>Tottori, Japan 1956 - 1984</td>
<td>Livebirths in Tottori prefecture of Honshu island, 650000 in population and averaging 8000 births per year.</td>
<td>Cases were ascertainment from multiple public hospitals and agency records confirmed by neuropediatricians at over 3 years of age. Deaths up to 3 years of age were included if CP known to be present. Postnatal cases were excluded.</td>
</tr>
<tr>
<td>Topp et al. (1997)</td>
<td>Eastern part of Denmark 1971 - 1986</td>
<td>All livebirths in eastern Denmark; these cover 50% of all births in Denmark, about 30000 births a year.</td>
<td>Since 1967, cases were collected by Danish CP Register from multiple sources. From birth year 1979, the data were added with a national register on hospital discharges. The cases were confirmed by at least 5 years follow up. Deaths before one year of age were excluded.</td>
</tr>
</tbody>
</table>
Table 3.3. Time trends in birthweight specified prevalence of cerebral palsy per 1000 livebirths.

<table>
<thead>
<tr>
<th>Authors (year)</th>
<th>Period of study</th>
<th>All weight</th>
<th>≤1000g</th>
<th>1001-1500g</th>
<th>1501-2000g</th>
<th>2001-2500g</th>
<th>&gt;2500g</th>
<th>Low birthweight (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dowding and Barry (1988)</td>
<td>1976 - 78</td>
<td>1.56</td>
<td>4.80</td>
<td>24.70</td>
<td>12.60</td>
<td>4.90</td>
<td>1.02</td>
<td>28.3% (≥3000g) 31.7%</td>
</tr>
<tr>
<td></td>
<td>1979 - 81</td>
<td>1.89</td>
<td>20.20</td>
<td>46.70</td>
<td>15.70</td>
<td>3.20</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hagberg et al. (1975, 1979, 1984, 1989, 1993, 1996)</td>
<td>1959 - 62</td>
<td>1.87</td>
<td></td>
<td>68.71 b1, b2</td>
<td>30.31</td>
<td>8.11</td>
<td>1.07</td>
<td>44.0%</td>
</tr>
<tr>
<td></td>
<td>1963 - 66</td>
<td>1.67</td>
<td></td>
<td>45.25 b1, b2</td>
<td>18.79</td>
<td>4.79</td>
<td>1.06</td>
<td>28.0%</td>
</tr>
<tr>
<td></td>
<td>1967 - 70</td>
<td>1.41</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>1971 - 74</td>
<td>1.58</td>
<td></td>
<td>60.24 b1, b2</td>
<td>20.74</td>
<td>6.19</td>
<td>1.12</td>
<td></td>
</tr>
<tr>
<td></td>
<td>1975 - 78</td>
<td>2.04</td>
<td></td>
<td>54.26 b1, b2</td>
<td>33.12</td>
<td>12.39</td>
<td>1.32</td>
<td></td>
</tr>
<tr>
<td></td>
<td>1979 - 82</td>
<td>2.17</td>
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<td>b4</td>
<td>b4</td>
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<td>1983 - 86</td>
<td>2.49</td>
<td>49.80 b2</td>
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<td>1987 - 90</td>
<td>2.36</td>
<td>56.60 b5</td>
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<td>Kavcic and Perat (1998)</td>
<td>1981 - 82</td>
<td>3.28</td>
<td></td>
<td>62.50</td>
<td>21.80</td>
<td>1.50</td>
<td>52.4%</td>
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<td>1983 - 84</td>
<td>3.39</td>
<td></td>
<td>49.40</td>
<td>17.30</td>
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<td>38.2%</td>
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<td></td>
<td>1985 - 86</td>
<td>3.06</td>
<td></td>
<td>43.30</td>
<td>16.60</td>
<td>1.50</td>
<td>43.6%</td>
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<td></td>
<td>1987 - 88</td>
<td>2.58</td>
<td></td>
<td>37.20</td>
<td>14.90</td>
<td>1.30</td>
<td>41.9%</td>
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<td></td>
<td>1989 - 90</td>
<td>2.45</td>
<td></td>
<td>32.70</td>
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<td>Meberg (1990) and Broch (1995)</td>
<td>1970 - 79</td>
<td>2.70</td>
<td></td>
<td>65.70</td>
<td>15.90</td>
<td>1.80</td>
<td>36.0%</td>
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<td></td>
<td>1980 - 89</td>
<td>2.10</td>
<td></td>
<td>44.40</td>
<td>18.40</td>
<td>1.30</td>
<td>41.0%</td>
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<td>1.42</td>
<td></td>
<td>17.11</td>
<td>5.52</td>
<td>1.25</td>
<td>30.4%</td>
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<td></td>
<td>1972 - 77</td>
<td>1.68</td>
<td></td>
<td>10.44</td>
<td>8.02</td>
<td>1.28</td>
<td>26.6%</td>
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<td></td>
<td>1978 - 83</td>
<td>2.21</td>
<td></td>
<td>55.95</td>
<td>11.16</td>
<td>1.24</td>
<td>47.4%</td>
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<tr>
<td></td>
<td>1984 - 89</td>
<td>c</td>
<td>c</td>
<td>c</td>
<td>c</td>
<td>c</td>
<td>48.0%</td>
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<tr>
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<td>1968 - 72</td>
<td>1.60</td>
<td>d</td>
<td>d</td>
<td>d</td>
<td>28.6%</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>1978 - 82</td>
<td>2.50</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>50.0%</td>
<td></td>
</tr>
<tr>
<td></td>
<td>1972 - 74</td>
<td></td>
<td>18.30</td>
<td>24.40</td>
<td>7.40</td>
<td>34.7%</td>
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<tr>
<td></td>
<td>1975 - 78</td>
<td>1.92</td>
<td>8.00</td>
<td>20.10</td>
<td>31.10</td>
<td>6.60</td>
<td>28.9%</td>
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<td></td>
<td>1979 - 82</td>
<td>1.89</td>
<td>26.20</td>
<td>30.30</td>
<td>19.00</td>
<td>6.10</td>
<td>34.8%</td>
<td></td>
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<tr>
<td></td>
<td>1983 - 85</td>
<td>2.18</td>
<td>47.40</td>
<td>67.00</td>
<td>14.00</td>
<td>6.60</td>
<td>44.0%</td>
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<td>Takeshita et al. (1989)</td>
<td>1956 - 59</td>
<td>2.42</td>
<td>\</td>
<td></td>
<td></td>
<td></td>
<td>about 10%</td>
<td></td>
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<tr>
<td></td>
<td>1971 - 74</td>
<td>1.43</td>
<td>\</td>
<td></td>
<td></td>
<td></td>
<td>before 1960</td>
<td></td>
</tr>
<tr>
<td></td>
<td>1975 - 80</td>
<td>0.57</td>
<td>\</td>
<td></td>
<td></td>
<td></td>
<td>to over 50%</td>
<td></td>
</tr>
<tr>
<td></td>
<td>1981 - 84</td>
<td>1.15</td>
<td>\</td>
<td></td>
<td></td>
<td></td>
<td>after 1980</td>
<td></td>
</tr>
<tr>
<td>Topp et al. (1997)</td>
<td>1971 - 74</td>
<td>1.70</td>
<td>fl</td>
<td>fl</td>
<td>fl</td>
<td>fl</td>
<td>fl</td>
<td></td>
</tr>
<tr>
<td></td>
<td>1975 - 78</td>
<td>1.60</td>
<td>fl</td>
<td>fl</td>
<td>fl</td>
<td>fl</td>
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<td></td>
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<tr>
<td></td>
<td>1979 - 82</td>
<td>2.30</td>
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<td>fl</td>
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<tr>
<td></td>
<td>1979 - 82f</td>
<td>2.60</td>
<td>fl</td>
<td>fl</td>
<td>fl</td>
<td>fl</td>
<td>fl</td>
<td></td>
</tr>
<tr>
<td></td>
<td>1982 - 86</td>
<td>3.00</td>
<td>fl</td>
<td>fl</td>
<td>fl</td>
<td>fl</td>
<td>fl</td>
<td>42.8%</td>
</tr>
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<td></td>
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<td></td>
<td></td>
<td></td>
<td></td>
<td>52.5%</td>
<td></td>
</tr>
</tbody>
</table>

31
Table 3.3. Time trends in birthweight specified prevalence of cerebral palsy per 1000 livebirths (cont).

<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>a</td>
<td>all data are prevalence of CP per 1000 births</td>
</tr>
<tr>
<td>a1</td>
<td>&gt;3000g prevalence per 1000 births</td>
</tr>
<tr>
<td>b</td>
<td>all data are incidence of CP per 1000 newborns surviving the first week of life.</td>
</tr>
<tr>
<td>b1</td>
<td>BW ≤ 1500g</td>
</tr>
<tr>
<td>b2</td>
<td>BW 1500-2499g</td>
</tr>
<tr>
<td>b3</td>
<td>BW 1500-2499g</td>
</tr>
<tr>
<td>b4</td>
<td>no definite data on the report</td>
</tr>
<tr>
<td>c</td>
<td>no population denominator data on the report</td>
</tr>
<tr>
<td>d</td>
<td>1968-72 &lt;2500g - 10.2%</td>
</tr>
<tr>
<td>e</td>
<td>incidence per 1000 livebirths born in 1971-1984</td>
</tr>
<tr>
<td>f</td>
<td>a national register on hospital discharges, founded in 1977, was used for supplementing the Danish Cerebral Palsy Register. The birth prevalence of CP in 1979-82 increased by 18%</td>
</tr>
<tr>
<td>f1</td>
<td>no prevalence data in birthweight subgroups</td>
</tr>
<tr>
<td>g</td>
<td>prevalence of preterm</td>
</tr>
</tbody>
</table>

1978-82 <2500g - 31.5%
and those under 1000g were very few. On the other hand, the perinatal mortality rate decreased proportionally across all gestations from 29 per 1000 births in 1954 to 5.6 in the period 1987-90 in the Southwest region of Sweden. Similarly, the neonatal mortality rate of infants weighing less than 1500g at birth fell from 67.5% in 1968-1971 to 28.7% in 1979-1981.

**Trends in birthweight specific prevalence**

Time trends in birthweight specific prevalence in each study are shown in Table 3.3. The crude prevalence of CP declined from the 1950s through the 1960s and then increased progressively through the 1970s in most studies, whereas those in Takeshita’s study significantly declined to their lowest prevalence during the period 1975-80 and increased thereafter. In contrast, the crude prevalence declined from 2.7 in 1970-79 to 2.1 in 1980-89 in Meberg’s study; and, similarly, the crude prevalence declined from 3.28 in 1981-82 to 2.45 in 1989-1990 in Slovenia. On the other hand, the crude prevalence did not significantly change from the 1950s through the 1980s in Western Australia. In the Swedish study, the data from 1963 through 1978 were calculated with birthweight specific incidence of CP per 1000 newborns surviving the first week of life. As there was a high perinatal mortality rate of low birthweight infants during that period, these data must be much higher than the data using livebirth as the denominator population. The increasing prevalence in low birthweight CP in their two later study periods was compatible with other reports, except for Meberg’s, and Kavcic and Perat’s. Most increasing trends were found in the very low birthweight group. Similarly, trends in Pharoah’s study showed that the lower the birthweight group, the later the rise in prevalence of CP occurred.
Trends in etiological groups

Hagberg et al. (1984, 1989) suggest that during the 1960s and 1970s, with respect to pathogenesis, the corresponding changes in CP incidence were mainly accounted for by the group with potential perinatal risk factors. The major changes occurred among preterm cases independent of etiology and among term cases with perinatal risk factors. Better diagnostic techniques such as ultrasound, CT and/or MRI are probably responsible for the recorded increase in the prenatal origin of CP.

In Hagberg’s latest report (1996), the single most frequent criterion for a prenatal etiology was based on CT findings of periventricular atrophy in 14 children (31%) who were born at term or near term in an uneventful delivery and with a normal neonatal history. The CT abnormalities were compatible with periventricular leukomalacias (PVL) interpreted as having occurred early in the third trimester. In addition, neuronal migration disorders of different kinds were diagnosed in 10 children (22%). The most frequent single criterion for a peri/neonatal etiology was intracerebral hemorrhage/stroke recorded in a total 32 children, 26 born preterm and six born at term. Hypoxic-ischemic encephalopathy not combined with intracerebral hemorrhage/stroke was present in 22 children, 11% of total cases; two born near term and 20 born at term. Moreover, in Takeshita’s study (1989), children with quadriplegia were found to have retarded intrauterine growth when compared with the normal population (P <0.05). It was considered that the damage to patients with quadriplegia might have started in the fetal period.

Trends in clinical type of CP

In most studies, diplegic CP predominated in the preterm group, whereas hemiplegic CP was more frequently found in the term group. For instance, in Hagberg’s study during the birth year period 1987-90, diplegia was present in 80% of the extremely preterm group, 66% in the
very preterm group, 58% in the moderately preterm group and 29% in the term group, while hemiplegia was present in 10, 16, 34, and 44% respectively. In contrast, in Western Australia in 1983-85, the main CP syndromes contributing to the rise in low birthweight infants with CP were spastic hemiplegia and quadriplegia. Similarly, the changing trends in clinical type of CP according to birthweight group in Pharoah’s 24 years study period showed that in the ≤ 1500g group there had been a highly significant fall in the proportion of all children with CP that were diplegic, from 57% in 1966-71 to 34% in 1984-89, and a concomitant increase in hemiplegia, from 13% to 33%. No striking change was found in the pattern in the 1501-2500g group; whereas in the > 2500g group the hemiplegias significantly increased, from 30% to 42%, with a concomitant fall in diplegias, from 16% to 10%, and in quadriplegias, from 43% to 36%.

**Trends in severity of CP**

Some studies evaluated the severity of the disability in a child with CP by assessing mental retardation, the ability to walk, and the presence of accompanying epilepsy; other studies did so by assessing severe learning, manual and ambulatory disability. In some of these studies, the severity of CP was relatively predominant in the preterm group or low birth weight group, and the proportion of severe cases also increased in the latest study period. For instance, in the earliest period of Pharoah’s study, 1966-71, the proportion of severely disabled in the normal birthweight group (> 2500g) was about twice that in the lowest birthweight group (≤ 1500g); whereas in the final study quarter, 1984-89, the proportion of severely disabled was very similar for all disability categories and birthweight groups. This changing trend occurred because there had been an increase in severity of disability in the very low birth weight groups but little change in those of normal birthweight. However, Hagberg’s latest study, which
covered the period from 1987 to 1990, reported a changing pattern among infants with an apparent decrease in the severity of disability among preterms (Hagberg et al., 1996).

3.4. Discussion

All these studies of prevalence of CP were performed in industrialized countries, mostly in Europe. Because all included studies were published in English, language bias and publication bias may exist. The criteria of applicability and validity used to assess the scientific quality of the articles, although based on criteria that are generally accepted in the current literature, may also be open to interpretation. The denominator population was taken to be all recorded livebirths whose mothers were resident in the study regions. Unavoidable prevalence-incidence bias may also have occurred because, although most cases were ascertained from multiple sources, failure of ascertainment may have occurred due to death or failure to follow up before a diagnosis was made. Failure to follow up was usually due to migration, but in most studies the cases which migrated in or out were respectively excluded or included from the denominator population.

The crude livebirth prevalence of CP from the 1950s through 1990 can be divided into two distinct trend eras: first, a significantly decreasing one during the 1950s and 1960s, a period when perinatal care became better organized and good basic methods of treatment were established; and second, a significantly increasing one during the 1970s and 1980s, a period which saw the development of mechanical ventilation and modern neonatal intensive care resulting in an increasing CP incidence related to improved survival of low birthweight infants. However, Hagberg’s latest study indicates a break (in the period 1987-90) in the gradual but significant increase in CP prevalence. Meanwhile, a declining population prevalence of CP in the 1980s was reported by Meberg and Broch in Norway (1995) and by Kavcic and Perat in
Slovenia (1998); where, according to the authors, there is a lag in the development of modern neonatal intensive care.

The prevalence of CP in low birthweight infants increased from the 1970s through the early 1980s in all these studies except in Meberg’s. The consistency of this observation in different countries strongly suggests that the trends are real. This increase occurred partly because recently developed intensive perinatal care led to a continuous decline in perinatal mortality and an increase in the survival of low birthweight infants. In Slovenia, it is noteworthy that a decreasing trend was observed in children born at \( \leq 38 \) weeks of gestation and in children with a birthweight of \(<1500\text{g}\) from 1981 to 1990. Some of this decrease may be attributable to several changes in perinatal care commenced in 1985 and to the introduction of neonatal intensive care in the tertiary maternity hospital in 1986.

There has been a sharp increase in multiple births, and the prevalence of CP in these births is greater than in singletons, the risk rising sharply with the number of babies, particularly among those with low birthweight and short gestation (Scheller and Nelson 1992; Petterson et al., 1993; MacGillivray and Campbell, 1995). This may be explained by the strong association of infertility treatment with increases in the prevalence of multiple births (Levene et al., 1992; Mordel et al., 1992). Another interesting observation is that, in a epidemiological study focusing on bilateral spastic cerebral palsy (defined as spasticity of the extremities on both sides, including diplegic, tetraplegic, and dyskinetic spastic factors) in Sweden and Germany (Krageloh-Mann et al., 1994), the preterm CP rate decreased after 1983, in spite of a continued fall in perinatal mortality. This finding is similar to Topp’s (1997) and Kavcic and Perat’s (1998) studies. It would seem to indicate that continuing improvements in perinatal care of very preterm infants helps them survive and also protects their immature brains against perinatal damage.
At present, there is only one published study of changes in the prevalence of CP in a population-based cohort during the 1990s (O'Shea et al., 1997, 1998). O'Shea's 1998 study reports that the decline in mortality among very low birth weight infants in the late 1980s and early 1990s has not resulted in an increase in the prevalence of CP. Another report by the same author (1997) suggests that the increasing survival of extremely low birthweight neonates (501 to 800 gm), from 20% to 59% between 1979 and 1994, has not resulted in an increased rate of major developmental problems identifiable at one year of adjusted age. O'Shea's studies may have several possible limitations such as selection bias, incomplete follow-up at 1 year of age, masked association between surfactant and the risk of CP (85% of subjects received surfactant therapy, all in third study epoch, 1989-1994), and the possibility of changing criteria for the diagnosis of CP during the study period. However, the finding that, since the 1980s, increasing survival rates of extremely low birthweight infants have not been accompanied by increasing rates of developmental problems also agrees with other previous studies (Robertson et al., 1992; La Pine et al., 1995; Kitchen et al., 1991; Tudehope et al., 1995; Saigal et al., 1989; Hack et al., 1996). The increased rate of neonatal survival and the decreased risk of CP, after the introduction of surfactant, imply that a decrease in brain damage attributable to complications of prematurity has occurred (Hamvas et al., 1996; Palta et al., 1994; Schwartz et al., 1994). It is also noteworthy that the incidence of CP in children born to those mothers who were preeclamptic or toxemic and who had received magnesium sulfate was less than for a comparable group of untreated mother (Kuban et al., 1992; Nelson and Grether, 1995). These observations need confirmation with carefully controlled clinical trials.

In a study focusing on CP in low birthweight infants, Powell and his colleagues concluded that factors influencing the rate of fetal development may be implicated in the etiology of diplegia (Powell et al., 1988b). Alternatively, various factors known at the time of
birth correctly identified most children as having a higher or lower estimated probability of hemiplegia (Powell et al., 1988a). Diplegic CP characterized the large majority of preterm cases. PVL is well known to be the major pathological cause of classical Little’s disease among infants with CP born moderately preterm. Volpe concluded that PVL is caused primarily by systemic hypotension, including episodes of disturbance so slight that they escape detection by even careful monitoring (Volpe, 1990). When all the preterm and term infants with CP are pooled together, adverse events during a period corresponding to 26-34 weeks of brain development are indicated as the prevailing causal factors. CNS destructive processes during gestational age 26-34 weeks, a period of particular vulnerability for periventricular brain structures, might account for more than half of the cases of CP in term infants, and three-quarters in preterm infants (Hagberg and Hagberg, 1993).

In analyzing the origin of CP, endeavours have been made to ascribe relative proportions to pre- and peripartum factors in the etiology of CP. Compared with older studies, the most recent reports have been more informative for tracing the timing of adverse events. This is mainly due to the successive introduction of ultrasound, CT, MRI and other neuroimaging techniques outside the regional neonatal centres. The information generated by these techniques allows more precise allocation to either a pre or perinatal group of origin. Generally, among children with CP born preterm, perinatal origin is predominant; whereas among children with CP born at term, prenatal origin is predominant. Some studies reported that the most frequent criterion for a prenatal etiology, based on the CT and/or MRI finding, is periventricular leukomalacia or atrophy interpreted as having occurred early in the third trimester (Krageloh-Mann et al., 1995; Sugimoto et al., 1995; Fazzi et al., 1994; Fujimoto et al., 1994; Rogers et al., 1994); whereas the most frequent criterion for a peri/neonatal etiology is intracerebral hemorrhage/stroke (Wiklund and Uvebrant, 1991; Wiklund et al., 1991). These
findings are compatible with Hagberg’s latest report (1996). On the other hand, the proportion of CP associated with intrapartum asphyxia in births in the United States was in the range of 3% to 13% (Nelson, 1988). Similar results, 8-9%, were reported in the Australian (Blair and Stanley, 1988,1993) and Swedish studies (Hagberg et al., 1996). All of these studies found that perinatal asphyxia is seldom the cause of CP.

A recent report (Olsen et al., 1997) regarding MRI studies of PVL and its clinical correlation in children found that the prevalence of PVL among all children born prematurely was 32%. It was observed in all children with CP, in 25% with minor neurological dysfunction, and in 25% of the clinically healthy preterm children. None of the children born at term had evidence of PVL. Children with PVL demonstrated poor performance on heel walking and Fog’s test. This was compatible with the predominant prevalence of diplegia in preterm cases.

The observed trends in the prevalence of CP (trends which were increasing in the 1970s and early 1980s) seem to be in decline in the late 1980s and early 1990s. This may be attributable to a combination of advanced diagnostic techniques, new biological and genetic tools (e.g. amniocentesis for chromosomal analysis, culture of fetal cells to detect certain metabolic derangements, and detection of neural-tube defects through maternal alpha-fetoprotein analysis), improved perinatal care, and modern intensive neonatal care. More investigation of the causative factors and the continued observation of the birth-weight specific epidemiology of CP will help to determine to what extent the syndrome has a prenatal origin and how much is attributable to perinatal factors. Correct attribution of prenatal or perinatal factors might also provide the opportunity to influence future trends in the prevalence of children with CP. Furthermore, a better understanding of changes in the prevalence of CP may give new clues about etiology which will be helpful in the prevention of CP.
Although a number of studies relating to the prevalence of CP have been published, there are methodological concerns about the current literature. Firstly, some researchers calculated prevalence of CP by using all births or neonatal survivors in the region as the denominator. This approach results in relative lower or higher prevalence as compared with studies using livebirths as the denominator population.

An additional difficulty in evaluating these papers is the lack of a standardized protocol. Researchers working in different countries tend to use slightly different definitions and slightly different criteria of inclusion and ascertainment. For instance, the duration of follow-up before a child was diagnosed as a case varied from 3 years to 5 years; postnatal or postneonatal cases were excluded in most studies, except in Hagberg’s serial reports and Riikonen’s study; inclusion or exclusion of deaths, or even the time of death, was different in most studies; and children with CP who migrated into or out of the study area during the study period were excluded in Kavcic and Perat’s study. A standardized protocol with very precise definitions, clear criteria of inclusion, and consistent ascertainment is required. Such a protocol would greatly increase the value of future studies.
Chapter 4. The survival of cerebral palsy: a review of population based studies

4.1. Introduction

During the past three decades, improvements in neonatal intensive care have dramatically reduced neonatal mortality among children of low birth weight (birth weight < 2500g), very low birth weight (birth weight <1500g), and extremely low birth weight (birth weight <1000g) (Pharoah et al., 1990; Naulty et al., 1994; Hagberg et al., 1993; Stanley and Watson, 1992; Kitchen et al., 1991; Hack and Fanaroff, 1990; Hoffman and Bennett, 1990; Kilbride et al., 1990; Riikonen et al., 1989) This has resulted in an increasing prevalence of cerebral palsy (CP) and changing clinical patterns of CP, as well as influencing the lifespan of patients with CP. While there are a number of studies examining the changing trends in rates of CP, information about survival of persons with CP is still limited (Tudehope et al., 1995; Hagberg et al., 1989; Saigal et al., 1989; Browen et al., 1993; Pharoah et al., 1996; Ens-Dokkum et al., 1994).

In addition to improved neonatal care, a marked improvement in the care of children with CP has occurred over the last 30 years. Children have access to improved health care and new technologies which allow them to have a better quality of life and fewer complications. Meanwhile social support systems allow the children to be cared for at home by their parents, by associated caregivers, or by foster parents (Miller and Eyman, 1978; Eyman et al., 1988; McCurley et al., 1972; Bilsard et al., 1988). As a result, most children with CP live in their communities and not in institutions.

While a number of studies (Balakrishnan and Wolf, 1976; Carter and Jancar, 1983; Chaney et al., 1979; Eyman et al., 1993) have addressed the survival of handicapped or mentally
retarded people, there is very little information about the life expectancy and associated problems of persons with CP. Such information would be useful for counseling parents and helping them to plan for the care of their children. Health, education, social service and community planners also need accurate life expectancy information to ensure that appropriate services are available. Increasingly, too, there are ethical questions raised about quality of life issues, the aggressiveness of "rescue" efforts (especially in the newborn period), and the rationing of services to certain segments of the population (Whitfield et al., 1997; Doyle et al., 1997; Sauve et al., 1998). The life expectancy of mobile individuals with CP and the possible use of survival statistics to help support litigation have also been a concern (Hall, 1995; Crichton, 1995; Miles, 1995). These are all significant issues and it is important to have accurate information about survival when dealing with them.

Although only a few papers reporting life expectancy of persons with CP have been published (Von Wendt et al., 1985; Kudrjavcev et al., 1985; Evans et al., 1990; Hutton et al., 1994; Crichton et al., 1995; Eyman et al., 1990, 1993; Strauss et al., 1998), there is a wide variety of opinion about the findings because of differences in internal variables such as study populations, methods of data collection, definitions and categories of study subjects, and additional diagnostic labels (Eyman et al., 1995; Hall, 1995; Crichton, 1995; Miles, 1995; Anderson, 1996; Strauss, 1997; Newton, 1997). The purpose of this chapter is to apply qualitative systematic review methods to these articles, and to estimate survival rates and prognostic factors for CP.

4.2. Data Sources and Methods

Several computerized bibliographic databases including MEDLINE, EMBASE, CINAHL, HealthSTAR, Biological Abstracts, and Current Contents in OVID Database were
utilized to search for articles. Search terms used were CP and survival, CP and life expectancy, and CP and mortality. Furthermore, other key words such as multihandicapped, multidisabled, and neurologically impaired were used instead of CP to search for articles through these databases.

Publications were included in this review if they examined population-based cohorts and reported survival or life expectancy data of persons with CP during the period 1966 to 1998. Editorial, commentaries, letters, unpublished studies, abstracts, reports appearing only in government publications, and summaries of presentations given at medical meetings or brief transcripts of meetings were excluded. Only data provided in the reports were considered, and no attempt was made to contact the authors for missing data. Only articles describing population-based registries of CP in which attempts were made to ascertain all individuals with CP in a region were included. Hospital-based studies were not included unless medical care of all persons with CP in the region was centralized at the regional hospital.

The scientific quality of the selected articles was assessed according to criteria of applicability and validity developed by Oxman and Guyatt (1988), Oxman et al. (1991), and Mulow (1987)—the same standards used to determined the quality of population-based studies of prevalence in Chapter 3. The elements for evaluating applicability were modified to focus mainly on specified purpose, definitions, criteria of inclusion and exclusion, ascertainment of case, sample size, methods of statistical analysis, and summary or conclusion. Validity was evaluated for elements such as play of chance, bias, and confounding.

Because there were so few articles compatible with our search criteria, we performed a narrative, qualitative summary of each study. We also reviewed other articles, literature and comments to discuss the issues relating to survival of patients with CP. Major textbooks for
relevant disciplines were also reviewed (Russman, 1998; Harris, 1995; Nelson et al., 1994; Pellegrino and Dormans, 1998).

4.3. Results

There were only five articles that qualified as presenting a population-based survival analysis of people with CP (Kudrjavcev et al., 1985; Evans et al., 1990; Hutton et al., 1994; Crichton et al., 1995; Strauss et al., 1998). These are presented in Table 4.1. Some overlap exists between different databases; therefore the number of cases unique to any given database is given in brackets in Table one. One of the 5, Kudrjavcev’s study (1995), although compatible with our search criteria, was not included in this paper because its sample size (total of 64 cases) was too small for meaningful survival analysis.

Characteristics of the four remaining reports are shown in Table 4.2. These studies were performed in different areas of the world including the United Kingdom, Canada, and the United States. The duration of each study ranged from 10 to 38 years with most subjects followed for at least 5 years, except those in Strauss’ study (1998). The population base in each study was greater than 230,000. Only one report recorded the ethnic background of subjects (Crichton et al., 1995). Individuals with acquired CP were excluded from Hutton and Strauss’ studies. Evans and Crichton did not indicate whether or not they considered the nature of the CP.

The scientific quality of these articles was assessed according to the above stated criteria for applicability and validity (Table 4.3). All articles specified purpose of study; however, only two articles (Hutton et al., 1994; Crichton et al., 1995) clearly stated the definitions of study items and a third article only listed some of the definitions in an overall table (Strauss et al., 1998). Although all subjects were ascertained from multiple sources in all studies and most cases were reported by clinicians, the absence of clear consistent definitions in Evans and
Strauss's papers diminished their usefulness for comparative purposes. Criteria of inclusion and exclusion were stated in three articles but not in Crichton's study. Cases were ascertained from registered multiple data sources in three studies (Evans et al., 1990; Hutton et al., 1994; Crichton et al., 1995), whereas the cases in Strauss' study were ascertained from all children with CP who received services from the California Department of Developmental Services through any one of 21 regional centres. The number of subjects in each study was 732, 1251, 3187, and 12719 respectively. The sample size of every study was large enough to perform statistical analysis. Different methods of survival analysis were utilized in each study. Evans' study performed survival analysis with two computer programs; the others used different computer software packages to conduct more complicated analyses including the Kaplan-Meier survival curve, Cox's proportional hazard model, and likelihood ratio tests (Hutton et al., 1994; Crichton et al., 1995; Strauss et al., 1998).

The validity of each study was evaluated for the possible influences of chance, bias, and confounding. To diminish the influence of chance, all of these population-based studies utilized statistical significance tests to determine the likelihood that sampling variability could explain the observed results, and they all established confidence intervals to indicate the range within which the true estimate of effect was likely to lie with a given degree of assurance. To avoid selection bias, all ascertained cases were evaluated or reported by pediatricians, pediatric neurologists or evaluation teams. All cases, except for Strauss', were followed for at least 5 years to confirm the diagnosis of CP. Strauss and colleagues, however, extracted data on all children diagnosed with CP between the age of 6 months and 3 years, 6 months. Their data therefore have some selection biases related to cases where the CP resolved or to cases where the nature of the developmental disabilities was progressively exacerbated. For example, in a review of data from the Collaborative Perinatal Project of the National Institute of Neurological
and Communicative Disorders and Stroke (Shapiro et al., 1987), 51% of children diagnosed as having CP at age 1 year were found to be free of motor handicap by 7 years of age. In Kudrjavcev’s study (1985), CP findings in eight children resolved before the seventh birthday—a resolution rate of 13% for the entire group of 64 cases. Finally, all studies performed stratified and multivariate analysis to control confounding.

Basic factors which might influence survival of people with CP in each study are shown in Table 4.4. There was no apparent difference in survival times between males and females in all studies. In Hutton’s study both birth weight and gestational age, which were not mentioned in Crichton’s report, were less predictive of survival than was functional disability. Similarly, birth weight had no important impact on survival in Evans and Strauss’s studies. While most of the deaths occurred in the quadriplegia group, the clinical type of CP was considerably less informative for prognostic purposes than the categories of functional disability in Evans, Hutton and Strauss’ studies. However, Crichton and colleagues reported that the survival in the group of hemiplegia / monoplegia was significantly better than that of those in the other groups, and that of the diplegia group was better than the quadriplegia group.

Direct comparison of the studies is complicated slightly by the fact that Evans established separate subgroups for “ataxia with or without spasticity” and for “dyskinetia with or without spasticity” rather than classifying such cases in the relevant spastic category. This classification results in his “other / unclassified” category making up 45% of total cases. Strauss’ study, meanwhile, included some ataxias, dyskinetias, and hypotonias in the quadriplegic group and consequently his results showed a high number of quadriplegias and a relatively lower mortality in the quadriplegic group (Fig. 4.1, Fig. 4.2).

Functional ability of persons with CP in each study is shown in table 4.5. Using multivariate analysis, Evans’ study showed that immobility and severe mental retardation were
the two strongest predictors of mortality. Hutton's study showed that ambulation was a better predictor of mortality than manual dexterity or mental ability and that manual dexterity was a better predictor than mental ability. Similarly, Strauss' study showed that the most powerful prognostic factors for survival were mobility and feeding skills. Whereas Crichton's study showed that type of CP, mental retardation, and epilepsy were independent prognostic factors of survival, there was no evidence to suggest that combinations of these factors improved or reduced survival chances.

Overall, deaths were generally higher among tube fed patients and concentrated in groups with the most severe disabilities such as severe ambulatory disability, severe manual disability, and severe mental retardation. In most studies the actual cause of death was not specified. In Evans' study, mention of a condition classifiable as CP appeared on 62% of death certificates, but only 22% of the death certificates had infantile cerebral palsy as the underlying cause of death; and in 78% of cases there was a respiratory (or cardiorespiratory) cause of death, most commonly a respiratory infection.

As did Evans' report, Hutton's study showed that the survival rate of mildly and even moderately affected children approached that of unaffected children, at least for the first 20 years. Even severely disabled children had about a 50% probability of surviving to 20 years of age. In Crichton's long term follow-up study (38 years), only 10 percent of the population died during the study period. The overall survival rate at 30 years was at least 87 percent. In Strauss' study, children with fair motor and eating skills had good survival prospects, with 90% or more reaching adulthood. However, among children who were unable to lift their head, median survival time was seven years for those who were tube fed and 14 years for those fed entirely by others.
Table 4.1. Number of citations and details of ineligible studies identified through different databases.

<table>
<thead>
<tr>
<th></th>
<th></th>
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<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Total number of citations</td>
<td>18(18)</td>
<td>13(1)</td>
<td>15(0)</td>
<td>6(0)</td>
<td>7(0)</td>
<td>2(0)</td>
<td>19</td>
</tr>
<tr>
<td>Total eligible studies</td>
<td>5(5)</td>
<td>4(0)</td>
<td>4(0)</td>
<td>3(0)</td>
<td>1(0)</td>
<td>1(0)</td>
<td>5</td>
</tr>
<tr>
<td>Total ineligible studies</td>
<td>13(13)</td>
<td>9(1)</td>
<td>11(0)</td>
<td>3(0)</td>
<td>6(0)</td>
<td>1(0)</td>
<td>14</td>
</tr>
<tr>
<td>1) Not a regional population based studies.</td>
<td>1(1)</td>
<td>0(0)</td>
<td>0(0)</td>
<td>0(0)</td>
<td>0(0)</td>
<td>0(0)</td>
<td>1</td>
</tr>
<tr>
<td>2) The population of the study was based on specified groups.</td>
<td>4(4)</td>
<td>3(0)</td>
<td>3(0)</td>
<td>2(0)</td>
<td>1(0)</td>
<td>1(0)</td>
<td>4</td>
</tr>
<tr>
<td>3) The studies were focused on acquired CP or on adults with CP.</td>
<td>1(1)</td>
<td>1(0)</td>
<td>1(0)</td>
<td>1(0)</td>
<td>1(0)</td>
<td>0(0)</td>
<td>1</td>
</tr>
<tr>
<td>4) Reports were editorials, letters, commentaries, unpublished studies, abstracts, and reviews.</td>
<td>7(7)</td>
<td>5(1)</td>
<td>7(0)</td>
<td>0(0)</td>
<td>4(0)</td>
<td>0(0)</td>
<td>8</td>
</tr>
<tr>
<td>5) Not written in English.</td>
<td>0(0)</td>
<td>0(0)</td>
<td>0(0)</td>
<td>0(0)</td>
<td>0(0)</td>
<td>0(0)</td>
<td>0</td>
</tr>
</tbody>
</table>

Note: 1. Obviously some studies show up in more than one database. The numbers in brackets indicate studies not found in the previously listed databases. Since MEDILINE is first listed, the number in brackets for MEDILINE is always identical to the total number of cases found.
2. The number in the “total” column is the total number of different studies identified through all databases.
3. No eligible citations were found using the Health and Psychosocial Instruments and the Cochrane Database of Systematic Reviews.
Table 4.2. Characteristics of studies on survival of persons with cerebral palsy (CP).

<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of Subjects</td>
<td>732</td>
<td>1251</td>
<td>3187</td>
<td>12719</td>
</tr>
<tr>
<td>Population</td>
<td>About 236,000 livebirths within the region between 1970-1974, (with 527 subjects with cerebral palsy).</td>
<td>Total population about 2.5 million, 25,000-30,000 births a year.</td>
<td>Total population of B.C. is about 3.5 million.</td>
<td>Total population of the State of California is about 32 million.</td>
</tr>
<tr>
<td>Ethnicity</td>
<td>was not mentioned</td>
<td>was not homogeneous, but was largely of European origin, with substantial Asian and native American populations.</td>
<td>was not mentioned but it was known to be not homogeneous.</td>
<td></td>
</tr>
<tr>
<td>Ascertainment of case</td>
<td>Initial ascertainment was from multiple sources, but later follow-up used the resources of the National Health Service Central Register.</td>
<td>A register of cases was compiled from multiple data sources to ensure completeness of ascertainment.</td>
<td>A population-based Health Surveillance Registry collected data from many sources.</td>
<td>The study included all children with CP who received any services from the California Department of Developmental Services through one of the 21 regional centres.</td>
</tr>
<tr>
<td>Method of Survival analysis</td>
<td>Survival analysis was done with computer programs BMDP1L and BMDP2L.</td>
<td>Tables of survival, log-logistic accelerated life model and likelihood ratio tests were used. The analyses were performed with the SAS software package.</td>
<td>Included Kaplan-Meier survival function plots, log rank test, and Cox's proportional hazard model. The statistical packages SPSS for Windows and EGRET were used to analyze the data.</td>
<td>Kaplan-Meier survival curve and Cox's proportional hazard model were used. All data were computerized using SAS with S-PLUS.</td>
</tr>
</tbody>
</table>
Table 4.3. The scientific quality of the studies.

<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Applicability</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Purpose of study specified</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Definitions stated</td>
<td>-</td>
<td>+</td>
<td>+</td>
<td>±</td>
</tr>
<tr>
<td>Criteria of inclusion and exclusion reported</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td>Ascertainment of cases from multiple sources</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Valid sample size</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Methods of statistical analysis stated</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Summary or conclusion reported</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td><strong>Validity</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Control of play of chance</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Avoidance of biases</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>±</td>
</tr>
<tr>
<td>Control of confounding</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
</tbody>
</table>

**Note:** "+" indicates that the authors addressed the relevant criteria.

"-" indicates failure to address criteria.

"±" indicates that while the authors attempted to address criteria they were not completely successful in doing so.
### Table 4.4. Basic factors influencing survival of persons with cerebral palsy.
Values are number of subjects (percentage) [number of deaths].

<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Sex</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Female</td>
<td>312 (43) [26]</td>
<td>541 (43) [55]</td>
<td>1454 (46) [142]</td>
<td>5570 (44) [565]</td>
</tr>
<tr>
<td>Male</td>
<td>420 (57) [47]</td>
<td>709 (57) [88]</td>
<td>1733 (54) [181]</td>
<td>7149 (56) [683]</td>
</tr>
<tr>
<td>Total</td>
<td>732 (100) [73]</td>
<td>1250 (100) [143]</td>
<td>3187(100)[323]</td>
<td>12719(100)[1248]</td>
</tr>
<tr>
<td><strong>Birth Weight (g)</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt;1500</td>
<td>birth weight has</td>
<td>134 (11) [7]</td>
<td>was not mentioned</td>
<td>\ 2512 (20) [154]</td>
</tr>
<tr>
<td>1501-2500</td>
<td>not any important</td>
<td>303 (26) [26]</td>
<td>/</td>
<td>/</td>
</tr>
<tr>
<td>&gt;2500</td>
<td>impact on survival</td>
<td>753 (63) [95]</td>
<td>/</td>
<td>10207 (80)[1094]</td>
</tr>
<tr>
<td>Total</td>
<td>1190(100)[128]</td>
<td>1190(100)[128]</td>
<td>/</td>
<td>12719(100)[1248]</td>
</tr>
<tr>
<td><strong>Type of cerebral palsy</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Quadriplegia</td>
<td>144 (20) [37]</td>
<td>424 (35)[108]</td>
<td>757 (25)[127]</td>
<td>7014 (55)[930]</td>
</tr>
<tr>
<td>Diplegia/paraplegia</td>
<td>100 (14) [0]</td>
<td>276 (23) [9]</td>
<td>412 (14) [19]</td>
<td>2140 (17) [86]</td>
</tr>
<tr>
<td>Other/unspecified</td>
<td>333 (45) [34]</td>
<td>128 (10) [11]</td>
<td>1003 (33) [124]</td>
<td>1672 (13) [177]</td>
</tr>
<tr>
<td>Total</td>
<td>732 (100) [73]</td>
<td>1225(100)[135]</td>
<td>3007(100)[303]</td>
<td>12719(100)[1248]</td>
</tr>
</tbody>
</table>
Fig. 4.1. Proportion of type of CP

- Evans et al.
- Hutton et al.
- Crichton et al.
- Strauss et al.

Legend:
- Red: Quadriplegia
- Green: Diplegia/paraplegia
- Blue: Hemiplegia/monoplegia
- Yellow: Other/unspecified
Fig. 4.2. Mortality in each type of CP

- Evans et al.
- Hutton et al.
- Crichton et al.
- Strauss et al.

Legend:
- Red: Quadriplegia
- Green: Diplegia/paraplegia
- Blue: Hemiplegia/monopleg
- Yellow: Other/unspecified
- Blue: Total

Percentage vs. CP Type
Table 4.5. Functional abilities influencing survival of persons with cerebral palsy. Values are number of subjects (percentage) [number of deaths].

<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Mental retardation</td>
<td>None</td>
<td>None</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td></td>
<td>Mild</td>
<td>Mild</td>
<td>Mild</td>
<td>Mild</td>
</tr>
<tr>
<td></td>
<td>Moderate</td>
<td>Moderate</td>
<td>Moderate</td>
<td>Moderate</td>
</tr>
<tr>
<td></td>
<td>Severe/Profound</td>
<td>Severe/Profound</td>
<td>Severe/Profound</td>
<td>Severe/Profound</td>
</tr>
<tr>
<td></td>
<td>Too young</td>
<td>Too young</td>
<td>Too young</td>
<td>Too young</td>
</tr>
<tr>
<td>Total</td>
<td>504 (100) [48]</td>
<td>1211 (100) [131]</td>
<td>3227 (100)[321]</td>
<td>12719 (100)[1248]</td>
</tr>
<tr>
<td>Mental retardation</td>
<td>None</td>
<td>None</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td></td>
<td>Mild</td>
<td>Mild</td>
<td>Mild</td>
<td>Mild</td>
</tr>
<tr>
<td></td>
<td>Moderate</td>
<td>Moderate</td>
<td>Moderate</td>
<td>Moderate</td>
</tr>
<tr>
<td></td>
<td>Severe/Profound</td>
<td>Severe/Profound</td>
<td>Severe/Profound</td>
<td>Severe/Profound</td>
</tr>
<tr>
<td></td>
<td>Too young</td>
<td>Too young</td>
<td>Too young</td>
<td>Too young</td>
</tr>
<tr>
<td>Total</td>
<td>504 (100) [48]</td>
<td>1211 (100) [131]</td>
<td>3227 (100)[321]</td>
<td>12719 (100)[1248]</td>
</tr>
<tr>
<td>Epilepsy</td>
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<td>Non-existent</td>
<td>Non-existent</td>
<td>Non-existent</td>
</tr>
<tr>
<td></td>
<td>Generalized</td>
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<tr>
<td></td>
<td>Partial</td>
<td>Partial</td>
<td>Partial</td>
<td>Partial</td>
</tr>
<tr>
<td></td>
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<td></td>
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</tr>
<tr>
<td>Ambulation</td>
<td>Minimal disability</td>
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<td>Minimal disability</td>
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</tr>
<tr>
<td></td>
<td>Limited mobility</td>
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<td>Limited mobility</td>
</tr>
<tr>
<td></td>
<td>Walking aids</td>
<td>Walking aids</td>
<td>Walking aids</td>
<td>Walking aids</td>
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<td></td>
<td>Self propelled</td>
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<td>Self propelled</td>
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<tr>
<td></td>
<td>wheelchair</td>
<td>wheelchair</td>
<td>wheelchair</td>
<td>wheelchair</td>
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<tr>
<td></td>
<td>Other wheelchair</td>
<td>Other wheelchair</td>
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<td>Other wheelchair</td>
</tr>
<tr>
<td></td>
<td>immobility</td>
<td>immobility</td>
<td>immobility</td>
<td>immobility</td>
</tr>
<tr>
<td>Unknown</td>
<td>124 [17] [31]</td>
<td>274 (22) [115]</td>
<td>124 [17] [31]</td>
<td>274 (22) [115]</td>
</tr>
<tr>
<td>Total</td>
<td>732 (100) [74]</td>
<td>1221 (100) [134]</td>
<td>732 (100) [74]</td>
<td>1221 (100) [134]</td>
</tr>
<tr>
<td>Manual dexterity</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td></td>
<td>Mild disability</td>
<td>Mild disability</td>
<td>Mild disability</td>
<td>Mild disability</td>
</tr>
<tr>
<td></td>
<td>Moderate disability</td>
<td>Moderate disability</td>
<td>Moderate disability</td>
<td>Moderate disability</td>
</tr>
<tr>
<td></td>
<td>Severe disability</td>
<td>Severe disability</td>
<td>Severe disability</td>
<td>Severe disability</td>
</tr>
<tr>
<td>Manual dexterity</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td></td>
<td>Mild disability</td>
<td>Mild disability</td>
<td>Mild disability</td>
<td>Mild disability</td>
</tr>
<tr>
<td></td>
<td>Moderate disability</td>
<td>Moderate disability</td>
<td>Moderate disability</td>
<td>Moderate disability</td>
</tr>
<tr>
<td></td>
<td>Severe disability</td>
<td>Severe disability</td>
<td>Severe disability</td>
<td>Severe disability</td>
</tr>
<tr>
<td>Total</td>
<td>1212 (100) [133]</td>
<td>1212 (100) [133]</td>
<td>1212 (100) [133]</td>
<td>1212 (100) [133]</td>
</tr>
</tbody>
</table>
4.4. Discussion

Numerous studies relating to issues of CP, such as prevalence, epidemiological trends, etiology, diagnosis, and treatment, have been published in the past decades (Torfs et al., 1990; Freeman and Nelson, 1988; Armstrong, 1992; Koman et al., 1994; Evans and Alberman, 1991; Mutch et al., 1992). There are also numerous reports discussing the issues of developmental disability and mental retardation. However, there are only five population-based articles addressing survival of people with CP and only four of these studies are included in this paper.

As the subjects of all these studies were collected from a population-based registry, some systemic errors or biases exist. For instance, CP was usually diagnosed only after several months of age. Hence infants who died in the first few months of life were less likely to have been defined as "cases." This means that the total number of deaths reported as occurring in the first year of life was lower than was actually the case. Since some of the missing data related to children who died before there had been any assessment of their functional disability, these missing values are unlikely to be independent of the severity of disability. The unassessed children might reasonably be expected to include a higher percentage of severely affected children, and this may have led to an overestimation of life expectancy of the more severely disabled groups.

Strauss and colleagues (1998) measured survival time only from the time of the first CP evaluation. Since survival time is usually measured from birth, Strauss’ method resulted in apparently shortened survival time and also led to the inclusion of age at the first CP evaluation as a potential risk factor. Furthermore, suspected CP was assessed by an evaluation team contracted by the California Department of Developmental Services as part of an initial evaluation for receiving services. The database was believed to include nearly all people in the state with severe CP, as such people had extensive need of medical and other services.
Presumably many children with mild disability were not included, and this might partly explain why the proportion of quadriplegia was a relatively high 55% in Strauss' subjects.

Eyman and his colleagues (1990, 1993) did a major study of people with profound developmental disability and mental retardation who received services from the California Department of Developmental Services, the same database as Strauss' (1998). It is difficult to analyze Eyman's figures accurately or to compare them with the other four CP studies. Eyman defined his study group according to developmental disability and severe mental retardation, excluded improved subjects from study subgroups every year, and did not limit his subjects to persons with CP (only 21.5% and 20.9%, respectively, of the subjects were considered to have CP). Consequently, although Eyman et al. used the same database as Strauss, the life expectancy reported is shorter.

Williams and Alberman (1998) recently reported that 85% of children with severe four-limb involvement survived during a 9 to 15 year period of follow-up, and no deaths occurred in children known to have less than four-limb involvement. These findings are compatible with the results of Evans, Hutton, Crichton, and Strauss, as all four studies showed that there was little or no mortality in children with less than four-limb involvement. From this it would appear that quadriplegic children with severe functional disability are at risk before they reach adolescence. Williams and Alberman also reported that the survival of individuals with severe quadriplegia known to have an additional diagnostic label was lower, though not significantly, than those with no known label.

While O'Shea et al. (1998) reported that the increasing survival of very low birth weight infants (birth weights of 500 to 1500 g) in the 1980s and 1990s has not resulted in an increased prevalence of CP among survivors; and the increasing survival of extremely low birth weight neonates (birth weights of 501 to 800 g) since the late 1970s has not resulted in an increased
rate of major developmental problems identifiable at one year of age (1997). CP was found in 12% of extremely immature infants (gestational age was 26 weeks or less) and 8% of extremely small infants (birth weights of 800 g or less) in a quantitative review report (Lorenz et al., 1998). Similarly, the crude prevalence of CP in multiple birth babies is much higher than that of singletons (Pharoah and Cooke, 1996; Williams et al., 1996). However, the information of survival for these populations is still very limited.

The life expectancies of children with CP in these studies show that the survival rate of mildly and even moderately affected children approaches that of unaffected children, at least for the first 20 years. Even severely disabled children have about a 50% probability of surviving to 20 years of age. Since many health professionals and government agencies tend to think of persons with CP as having significantly reduced life expectancies, these results should prompt a reappraisal of the long term needs of persons with CP. Physicians should make parents aware of the long term prognosis of a child with CP, so that the parents can make realistic long term plans and commitments. Similarly, health, education, social and community planners should take these survival rates into account when budgeting for care, equipment and facilities for individuals with CP.

Overall, the results of these studies contribute important and valuable information about the survival of people with CP. However, there methodologic limitations exist. One of the difficulties in evaluating the above papers is the lack of a standardized protocol. Researchers working in different countries, sometimes even within the same country, tend to use different definitions, different criteria of inclusion and ascertainment, and different prognostic factors for survival. For instance, Evans and Hutton classified CP with different definitions. Strauss chose to classify ataxic, dyskinetic and hypotonic types of CP as quadriplegic, and consequently he had a high proportion of quadriplegics and a reduced quadriplegic mortality rate relative to the
other studies. Duration of follow-up and confirmation of diagnosis were other issues. All cases in the studies by Evans, Hutton, and Crichton were followed for at least 5 years to confirm the diagnosis of CP, whereas Strauss extracted data on all children diagnosed with CP between the age of 6 months and 3 years, 6 months. Persons with acquired CP were excluded from Hutton and Strauss' studies, whereas Evans and Crichton did not indicate the exclusion criteria. All studies used different items to evaluate prognostic factors.

Another issue is that information relating to the factors influencing survival of persons with CP, such as orthopaedic, visual, and hearing impairments, feeding problems, respiratory infection, and technical advances and improvements in the management of these problems, is still very limited. A standardized protocol with very precise definitions, clear criteria of inclusion, consistent ascertainment, and as complete consideration of influencing factors and variations as possible is needed and would greatly increase the comparability and usefulness of future studies.
Chapter 5. Prevalence and survival of children with cerebral palsy in British Columbia — development of data extraction form and pre-test study

5.1. Introduction

Cerebral palsy (CP) is a group of manifestations of impaired neurologic function caused by a non-progressive brain abnormality dating to events in the prenatal or perinatal periods. It is often associated with motor disability, mental retardation, epilepsy, and hearing and visual impairments. Although there has been marked improvement in the care of children with CP over the last 30 years, currently the information on prevalence and survival is limited. Health professionals need reasonably accurate information about life-expectancy and outcome of management so that they may provide better counseling for the families of those with CP, as well as better planning for their care, education and employment needs.

While a number of studies relating to the changing trends of the prevalence of CP have been reported in industrialized countries (Hagberg et al., 1996; Pharoah et al., 1996; Stanley and Watson, 1992; Topp et al., 1997; Kavcic and Perat, 1998), there is still very limited information on the prevalence of CP in British Columbia. Recently, a few studies focusing on survival of CP were done in the United Kingdom and the United States (Evans et al., 1990; Hutton et al., 1994; Strauss et al., 1998). The researchers used slightly different definitions, criteria of inclusion, ascertainment, and prognostic factors. This information is not easily comparable and cannot be readily be extrapolated to the situation in British Columbia (BC) and Canada.

Although some preliminary work focusing on survival and prognostic factors with type of CP, epilepsy and severity of mental retardation has been done in British Columbia (Crichton
et al., 1995), the findings are not sufficiently detailed to provide the information required about prognostic factors in maternal and prenatal conditions, neonatal complications, etiology of CP, motor and other functional disabilities, and respiratory, feeding and orthopaedic problems.

To evaluate the prevalence and survival of children with CP in BC, we propose a retrospective study of medical charts based on information identified through two databases. To extract the information in a consistent and meaningful form, a Cerebral Palsy Chart Review Protocol was developed. The usefulness of this protocol was assessed by running a pre-test on a convenience sample from a hospital. The necessary modifications were made to the Protocol. This revised Protocol will be used for the retrospective study.

5.2. Methods

A data extraction form, the Cerebral Palsy Chart Review Protocol (Appendix A), was designed for the purpose of reviewing the charts of patients with CP from hospitals or treatment-centre based records in BC. Because there are many differing types of CP with various associated clinical problems and many possible etiological pathways, it was felt that the protocol had to provide health professionals with as detailed and accurate a description of individual cases as possible. With this in mind the protocol was designed to screen the charts for descriptive data, prenatal information, newborn data, diagnosis, functional ability, surgical intervention, and cause of death.

The protocol surveys four main categories: general data, maternal record, newborn record, and CP record. Taken together these categories provide meaningful information about the prevalence of specific forms of CP and about the life expectancy with associated problems. General data consists of basic data such as birthdate, place of birth, gender, and family history. Maternal record mainly focuses on conditions or complications originating in the pregnancy, and
in the prenatal and perinatal periods. Newborn record includes the child’s condition at birth, neonatal disorders, and interventions. This information is valuable in understanding the etiological factors or the effective period of causative factors of CP.

The CP record consists of etiology, classification, severity, level of disability, mental ability, epilepsy, respiratory, feeding and orthopaedic problems, management, and outcome. The etiology items include prenatal, intranatal, postnatal, prematurity, and unclassified factors. The etiologic factors are important to assess the changing trends in prevalence of CP and to evaluate the effect of various interventions with respect to the prevention of CP.

CP is classified as spastic, dyskinetic, ataxic, atonic and mixed type. The spastic type is topographically subgrouped into monoplegia, hemiplegia, paraplegia, diplegia and quadriplegia. The different types of CP may relate to causative factors and may have a great influence on intervention and prognosis of CP.

Severity of CP is divided into five levels according to the Gross Motor Function Classification System (Russell et al., 1989; Russell et al., 1993; Palisano et al., 1997). For more detailed information of severity, the level of disability is subdivided into two parts: “motor domain” and “other functional domains.” We defined the motor domain according to parameters developed by the California Department of Developmental Services in their Client Development Evaluation Report (1978; 1986; Harris et al., 1982)

In addition to motor domain, level of disability also includes other functional domains such as eating skills, toilet training, receptive nonverbal ability, speech skills, language recognition skills and social response. This information is helpful in evaluating management and outcome of CP. Mental ability, type and response after treatment of epilepsy, respiratory, feeding and orthopaedic problems are other prognostic factors influencing the management and
outcome of CP. Length of survival is also influenced by medical treatment and/or surgical intervention (McGrath et al., 1992).

These items were included in the protocol because they all help to establish the type of CP, the etiological factors, the functional disabilities, and the prognostic factors. For ease of evaluation, all four main categories of data were assimilated in a data summary record.

To assess the feasibility of the Cerebral Palsy Chart Review Protocol, we conducted a pre-test on a convenience sample of charts from a hospital for children with disabilities. Thirty-five randomly selected charts of subjects with CP were reviewed. This pre-test study examined the type of information that can be abstracted from the hospital charts. In this pilot study, preliminary descriptive analyses were conducted to identify the characteristics of the sample by their survivorship status.

5.3. Results

In this pre-test, thirty five charts were randomly selected and reviewed using the Cerebral Palsy Chart Review Protocol. One was excluded because the symptoms of CP were caused by head injury suffered at 6 years of age, and therefore did not meet the definition of primary CP. Upon review, the charts did not provide complete information in all cases. The chart data relating to maternal pregnancy complications were not available in half the cases (17/34) and data of complication of labour were not found in 44% (15/34). Also, nearly one quarter (8/34) of newborn records were not available. The actual time of diagnosing an individual with CP was noted only in 18% of cases (6/34) and age of starting treatment or early intervention was often not available in most data charts. Data on both toilet training and epilepsy and outcome of epilepsy were incomplete in three cases.
Although the data was incomplete in some cases and although our sample size was too small to be statistically meaningful, the protocol allowed us to make the following observations about the charts reviewed. Two of the fifteen females (13%) and three of the nineteen males (16%) died (Table 5.1). In considering the etiological factors, 2 of the 9 with prenatal factors, 2 of the 3 with perinatal factors, and 1 of the 7 with postnatal factors died. One quarter of the total cases (8/34) were attributable to prematurity factors. The etiological factor was unclassifiable in nearly one fifth of total cases (6/34). More than three quarters of the cases (26/34), including all deaths, had spastic quadriplegia. Similarly, nearly three quarters (24/34), including all deaths, were severely mentally retarded. In addition, three quarters (25/34), including all deaths, were immobile or severely disabled.

Almost 60% of the cases (20/34) had visual impairment and 12% (4/34) had hearing impairment (Table 5.2). Four of the 18 cases with poor cognitive ability died (Table 5.3). Similarly, 4 of the 19 cases with poor social response died. Seven cases were unable to feed themselves; and fifteen cases, including all deaths, were tube fed. About two thirds of cases, including all deaths, were not toilet trained.

Only 15% of the cases (5/34) showed no symptoms of epilepsy (Table 5.4). About one third of epilepsy cases were partial epilepsy with secondary generalization. In these epileptic cases, one fifth still had frequent seizure on medication, and nearly two fifths had occasional seizures on medication.

One third (11/34) of the cases, including all deaths, had respiratory problems (Table 5.5); and aspiration pneumonia was documented in all deaths. Half of the cases (17/34), including all deaths, had feeding problems (Table 5.6). Fifteen cases were fed though gastrostomy and/or jejunostomy. Only two cases had no orthopaedic problems (Table 5.7). Twenty four of the 32 cases had spinal deformity, and 16 had subluxated or dislocated hips.
Nineteen cases, including all deaths, had received surgical interventions. One case had received intrathecal injection with Baclofen for severe spasticity.

Four of the five deceased patients died of respiratory distress and failure. One of these four also had induced sepsis and disseminated intravascular coagulation. The fifth patient died immediately following surgery for jejunostomy at hospital. Finally, the deceased cases were all associated with spastic quadriplegia, severe mental retardation, immobility, and also with feeding, respiratory and orthopaedic problems (Table 5.8).

As there was a limitation of sample size, we cannot have any further analysis and conclusion.
Table 5.1. Characteristics of individuals with cerebral palsy (N = 34).

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Death n (%)</th>
<th>Survivors n (%)</th>
<th>Total n</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Sex</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Female</td>
<td>2 (13.3)</td>
<td>13 (86.7)</td>
<td>15</td>
</tr>
<tr>
<td>Male</td>
<td>3 (15.8)</td>
<td>16 (84.2)</td>
<td>19</td>
</tr>
<tr>
<td><strong>Etiology of C. P.</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Prenatal factor</td>
<td>2 (22.2)</td>
<td>7 (77.8)</td>
<td>9</td>
</tr>
<tr>
<td>Perinatal factor</td>
<td>2 (66.7)</td>
<td>1 (33.3)</td>
<td>3</td>
</tr>
<tr>
<td>Postnatal factor</td>
<td>1 (14.3)</td>
<td>6 (85.7)</td>
<td>7</td>
</tr>
<tr>
<td>Prematurity factor</td>
<td>-</td>
<td>8 (100.0)</td>
<td>8</td>
</tr>
<tr>
<td>Unclassified factor</td>
<td>-</td>
<td>6 (100.0)</td>
<td>6</td>
</tr>
<tr>
<td>Incomplete data</td>
<td>-</td>
<td>1 (100.0)</td>
<td>1</td>
</tr>
<tr>
<td><strong>Type of C. P.</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Spastic quadriplegia</td>
<td>5 (19.2)</td>
<td>21 (77.8)</td>
<td>26</td>
</tr>
<tr>
<td>Spastic diplegia</td>
<td>-</td>
<td>1 (100.0)</td>
<td>1</td>
</tr>
<tr>
<td>Spastic hemiplegia</td>
<td>-</td>
<td>2 (100.0)</td>
<td>2</td>
</tr>
<tr>
<td>Atonic type</td>
<td>-</td>
<td>2 (100.0)</td>
<td>2</td>
</tr>
<tr>
<td>Mixed type</td>
<td>-</td>
<td>3 (100.0)</td>
<td>3</td>
</tr>
<tr>
<td><strong>Mental retardation</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>None</td>
<td>-</td>
<td>1 (100.0)</td>
<td>1</td>
</tr>
<tr>
<td>Moderate</td>
<td>-</td>
<td>9 (100.0)</td>
<td>9</td>
</tr>
<tr>
<td>Severe or suspected severe</td>
<td>5 (20.8)</td>
<td>19 (79.2)</td>
<td>24</td>
</tr>
<tr>
<td><strong>Severity of motor function (according to Gross Motor Function Classification System)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Level 3</td>
<td>-</td>
<td>7 (100.0)</td>
<td>7</td>
</tr>
<tr>
<td>Level 4</td>
<td>-</td>
<td>2 (100.0)</td>
<td>2</td>
</tr>
<tr>
<td>Level 5</td>
<td>5 (20.0)</td>
<td>20 (80.0)</td>
<td>25</td>
</tr>
</tbody>
</table>
Table 5.2. Visual and hearing impairments among individuals with cerebral palsy (N = 34).

<table>
<thead>
<tr>
<th></th>
<th>Deaths n (%)</th>
<th>Survivors n (%)</th>
<th>Total n</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Visual impairment</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cortical visual impairment</td>
<td>2 (11.8)</td>
<td>15 (88.2)</td>
<td>17</td>
</tr>
<tr>
<td>Optic nerve atrophy</td>
<td>-</td>
<td>2 (100.0)</td>
<td>2</td>
</tr>
<tr>
<td>Complicated visual impairment</td>
<td>1 (100.0)</td>
<td>-</td>
<td>1</td>
</tr>
<tr>
<td>No visual impairment</td>
<td>2 (14.3)</td>
<td>12 (85.7)</td>
<td>14</td>
</tr>
<tr>
<td><strong>Hearing impairment</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>One ear hearing impairment</td>
<td>1 (50.0)</td>
<td>1 (50.0)</td>
<td>2</td>
</tr>
<tr>
<td>Two ear hearing impairment</td>
<td>1 (50.0)</td>
<td>1 (50.0)</td>
<td>2</td>
</tr>
<tr>
<td>No hearing impairment</td>
<td>3 (10.0)</td>
<td>27 (90.0)</td>
<td>30</td>
</tr>
</tbody>
</table>
Table 5.3. Cognitive ability and daily living skills among individuals with cerebral palsy (N = 34).

<table>
<thead>
<tr>
<th></th>
<th>Deaths n (%)</th>
<th>Survivors n (%)</th>
<th>Total n</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Cognitive ability</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Well</td>
<td>-</td>
<td>1 (100.0)</td>
<td>1</td>
</tr>
<tr>
<td>Moderate</td>
<td>1 (6.7)</td>
<td>14 (93.3)</td>
<td>15</td>
</tr>
<tr>
<td>poor</td>
<td>4 (22.2)</td>
<td>14 (77.8)</td>
<td>18</td>
</tr>
<tr>
<td><strong>Social response</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Well</td>
<td>-</td>
<td>1 (100.0)</td>
<td>1</td>
</tr>
<tr>
<td>Moderate</td>
<td>1 (7.1)</td>
<td>13 (92.9)</td>
<td>14</td>
</tr>
<tr>
<td>poor</td>
<td>4 (21.1)</td>
<td>15 (78.9)</td>
<td>19</td>
</tr>
<tr>
<td><strong>Eating skill</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Finger feeding</td>
<td>-</td>
<td>12 (100.0)</td>
<td>12</td>
</tr>
<tr>
<td>Unable to feed by self</td>
<td>-</td>
<td>7 (100.0)</td>
<td>7</td>
</tr>
<tr>
<td>Tubular feeding</td>
<td>5 (33.3)</td>
<td>10 (66.7)</td>
<td>15</td>
</tr>
<tr>
<td><strong>Toilet train</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Trained</td>
<td>-</td>
<td>6 (100.0)</td>
<td>6</td>
</tr>
<tr>
<td>Partial trained</td>
<td>-</td>
<td>3 (100.0)</td>
<td>3</td>
</tr>
<tr>
<td>Not trained</td>
<td>5 (22.7)</td>
<td>17 (77.3)</td>
<td>22</td>
</tr>
<tr>
<td>Incomplete data</td>
<td>-</td>
<td>3 (100.0)</td>
<td>3</td>
</tr>
</tbody>
</table>
Table 5.4. Epilepsy and outcome of epilepsy among individuals with cerebral palsy (N = 34).

<table>
<thead>
<tr>
<th></th>
<th>Deaths n (%)</th>
<th>Survivors n (%)</th>
<th>Total n</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Epilepsy</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Generalized</td>
<td>1 (20.0)</td>
<td>4 (80.0)</td>
<td>5</td>
</tr>
<tr>
<td>Partial</td>
<td>1 (25.0)</td>
<td>3 (75.0)</td>
<td>4</td>
</tr>
<tr>
<td>Infantile spasm</td>
<td>-</td>
<td>3 (100.0)</td>
<td>3</td>
</tr>
<tr>
<td>Partial with 2nd generalization</td>
<td>1 (9.1)</td>
<td>10 (90.9)</td>
<td>11</td>
</tr>
<tr>
<td>Unclassified</td>
<td>-</td>
<td>3 (100.0)</td>
<td>3</td>
</tr>
<tr>
<td>Existent, with incomplete data</td>
<td>1 (33.3)</td>
<td>2 (66.7)</td>
<td>3</td>
</tr>
<tr>
<td>Non-existent</td>
<td>1 (20.0)</td>
<td>4 (80.0)</td>
<td>5</td>
</tr>
<tr>
<td><strong>Outcome of epilepsy</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Frequently seizure on medication</td>
<td>1 (14.3)</td>
<td>6 (85.7)</td>
<td>7</td>
</tr>
<tr>
<td>Occasionally seizure on medication</td>
<td>3 (23.1)</td>
<td>10 (76.9)</td>
<td>13</td>
</tr>
<tr>
<td>Seizure free on medication</td>
<td>-</td>
<td>2 (100.0)</td>
<td>2</td>
</tr>
<tr>
<td>Seizure free off medication</td>
<td>-</td>
<td>3 (100.0)</td>
<td>3</td>
</tr>
<tr>
<td>Incomplete data</td>
<td>-</td>
<td>4 (100.0)</td>
<td>4</td>
</tr>
</tbody>
</table>
Table 5.5. Respiratory problems among individuals with cerebral palsy (N = 34).

<table>
<thead>
<tr>
<th></th>
<th>Deaths n (%)</th>
<th>Survivors n (%)</th>
<th>Total n</th>
</tr>
</thead>
<tbody>
<tr>
<td>Existent</td>
<td>5 (45.5)</td>
<td>6 (54.5)</td>
<td>11</td>
</tr>
<tr>
<td>Non-existent</td>
<td>-</td>
<td>23 (100.0)</td>
<td>23</td>
</tr>
</tbody>
</table>

*All deceased cases had respiratory problems; only 6 of the 29 survivors had respiratory problems.*
Table 5.6. Feeding problems among individuals with cerebral palsy (N = 34).

<table>
<thead>
<tr>
<th></th>
<th>Deaths</th>
<th></th>
<th>Survivors</th>
<th></th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>n</td>
<td>(%)</td>
<td>n (%)</td>
<td></td>
<td>n</td>
</tr>
<tr>
<td><strong>Feeding problem</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Existent</td>
<td>5</td>
<td>(29.4)</td>
<td>12</td>
<td>(70.6)</td>
<td>17</td>
</tr>
<tr>
<td>Non-existent</td>
<td>-</td>
<td></td>
<td>17</td>
<td>(100.0)</td>
<td>17</td>
</tr>
<tr>
<td><strong>Management of feeding problem (n = 17)</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Gastrostomy</td>
<td>2</td>
<td>(18.2)</td>
<td>9</td>
<td>(81.8)</td>
<td>11</td>
</tr>
<tr>
<td>Gastrostomy + jejunostomy</td>
<td>2</td>
<td>(100.0)</td>
<td>-</td>
<td></td>
<td>2</td>
</tr>
<tr>
<td>Gastrostomy + fundoplication + jejunostomy</td>
<td>1</td>
<td>(50.0)</td>
<td>1</td>
<td>(50.0)</td>
<td>2</td>
</tr>
<tr>
<td>Saliva gland relocation</td>
<td>-</td>
<td></td>
<td>1</td>
<td>(100.0)</td>
<td>1</td>
</tr>
<tr>
<td>Treated with changing feeding position</td>
<td>-</td>
<td></td>
<td>1</td>
<td>(100.0)</td>
<td>1</td>
</tr>
</tbody>
</table>
Table 5.7. Orthopaedic problems among individuals with cerebral palsy (N = 34).

<table>
<thead>
<tr>
<th>Orthopaedic problem</th>
<th>Deaths n (%)</th>
<th>Survivors n (%)</th>
<th>Total n</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Existent</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Scoliosis or kyphosis or lordosis or kyphoscoliosis</td>
<td>5 (15.6)</td>
<td>27 (84.4)</td>
<td>32</td>
</tr>
<tr>
<td>Subluxated or dislocated hip</td>
<td>5 (20.8)</td>
<td>19 (79.2)</td>
<td>24</td>
</tr>
<tr>
<td>Non-existent</td>
<td></td>
<td>2 (100.0)</td>
<td>2</td>
</tr>
<tr>
<td><strong>Management of orthopaedic problem</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Supportive treatment</td>
<td>5 (15.6)</td>
<td>27 (84.8)</td>
<td>32</td>
</tr>
<tr>
<td>Surgical treatment</td>
<td>5 (26.3)</td>
<td>14 (73.7)</td>
<td>19</td>
</tr>
<tr>
<td>Intrathecal injection with Baclofen</td>
<td></td>
<td>1 (100.0)</td>
<td>1</td>
</tr>
</tbody>
</table>
Table 5.8. Proportion for each characteristic of deceased individuals with cerebral palsy.

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Deaths</th>
<th>Survivors</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>n</td>
<td>n (%)</td>
<td></td>
</tr>
<tr>
<td>Spastic quadriplegia</td>
<td>5 (19.2)</td>
<td>21 (80.8)</td>
<td>26</td>
</tr>
<tr>
<td>Severe mental retardation</td>
<td>5 (20.8)</td>
<td>19 (79.2)</td>
<td>24</td>
</tr>
<tr>
<td>Immobility</td>
<td>5 (20.0)</td>
<td>20 (80.0)</td>
<td>25</td>
</tr>
<tr>
<td>Tubular feeding</td>
<td>5 (33.3)</td>
<td>10 (66.7)</td>
<td>15</td>
</tr>
<tr>
<td>Not toilet trained</td>
<td>5 (22.7)</td>
<td>17 (77.3)</td>
<td>22</td>
</tr>
<tr>
<td>Existent respiratory problems</td>
<td>5 (45.5)</td>
<td>6 (54.5)</td>
<td>11</td>
</tr>
<tr>
<td>Documented aspiration pneumonia</td>
<td>5 (55.6)</td>
<td>4 (44.4)</td>
<td>9</td>
</tr>
<tr>
<td>Existent orthopaedic problems</td>
<td>5 (15.6)</td>
<td>27 (84.4)</td>
<td>32</td>
</tr>
<tr>
<td>Surgical therapy of orthopaedic problems</td>
<td>5 (26.3)</td>
<td>14 (73.7)</td>
<td>19</td>
</tr>
</tbody>
</table>
5.4. Discussion

We designed the Cerebral Palsy Chart Review Protocol to retrieve data from medical charts for information relating to etiological factors, functional abilities, and associated problems of CP. In addition to reviewing maternal and neonatal records, the Chart Review Protocol extracts information about etiology, classification, severity, level of disability, mental ability, epilepsy, respiratory, feeding and orthopaedic problems, management, and outcome. These items are designed to search for possible clues about etiologies or causative factors of CP, associated issues of CP, prognostic factors for CP, and prevention of CP. This information is important for epidemiological studies and for clinical practice.

After reviewing these sample charts, we found that, overall, the Chart Review Protocol allowed data to be extracted in a concise meaningful form. Variations in patient history and in the thoroughness of the records kept meant that the information collected varied in detail, but nevertheless there was enough information to help evaluate the factors which might influence survival of children with CP in BC. Because visual and hearing impairments were often noted in the charts and because knowledge of such impairments is of epidemiological or clinical importance we added sections on visual and hearing impairments. In the interests of concision or specificity of classification we also made modifications to some items such as neonatal record, level of disability, mental ability, and feeding and orthopaedic problems.

The data extraction form showed that less than one fifth of the medical charts established the actual point at which CP was diagnosed. Also, most data charts were unclear as to when treatment or early intervention was started. The lack of information in these areas may be due to the difficulty of having a confirmed diagnosis of CP in early infancy.

While the level of disability of CP is evaluated with various criteria in other reports, our protocol (Appendix A) and its revision (Appendix B) were designed to extract as much
significant information as possible about mobility, mental ability, and manual dexterity. Because the Client Development Evaluation Report evaluations of motor domain were difficult to assess, the level of disability in our revised protocol focused primarily on mobility, mental ability and manual dexterity, rather than on motor domain parameters. These three areas were felt to be most useful because they are comprehensive and practical, including as they do motor function, mobility skills, cognitive process, and daily living skills. Assessment of severity of CP was also revised to include level of disability and sensory impairment such as visual and hearing impairments.

The pre-test study was conducted on randomly selected charts from a hospital for children with disabilities. The use of such a hospital and the attendant selection bias probably means that a disproportionately high number of the patients were severely disabled. This is compatible with our result that three quarters of sample cases (25/34) were severely disabled. If the study had been conducted on charts from a general hospital, a hospital with a higher proportion of mild cases, the data might have been more difficult to extract since patients with mild CP do not require aggressive intervention and their condition is not scrutinized as exhaustively. The protocol may indeed be most effective for extracting information on patients with moderate or severe CP.

The pre-test showed that most data needed in our extraction form were retrievable from medical charts. However, data relating to pregnancy and maternal delivery were not available in about half of the cases, and newborn records were unavailable in about one quarter of the cases. When the data was combined with other information obtained from other records or reference papers on the charts, only one case had incomplete data on etiological factors. Insufficient information was mostly found when subjects were put into foster care or had migrated from foreign countries.
Through this pre-test study, we found this Chart Review Protocol to be a feasible extraction method. We therefore propose that the Cerebral Palsy Chart Review Protocol be used for a full scale study evaluating the prevalence of CP and survival of children with moderate to severe CP in British Columbia.
Chapter 6. Proposal for establishing the prevalence and survival of children with cerebral palsy in British Columbia

6.1. Introduction and Background

Cerebral palsy (CP) is the term used to describe a movement disorder thought to be the result of non-progressive brain dysfunction caused by disordered development or brain damage incurred during pregnancy, delivery, or in early life. CP is a symptom complex, rather than a specific disease. It is mainly a disorder of movement and posture with onset in the early years of life. The cerebral lesions of CP are static and nonprogressive; but changes do occur as the central nervous system matures, and peripheral physical symptoms may change with brain development. For instance, a hypotonic infant may become spastic or rigid, and a child who is originally diagnosed as choreoathetoid may subsequently become dystonic and develop contractures. CP is often associated with motor disability, mental retardation, epilepsy, and visual and hearing impairments.

During the past three decades, improvements in perinatal and neonatal intensive care have dramatically reduced the perinatal mortality among children of low birth weight (<2500g), very low birth weight (<1500g), and extremely low birth weight (<1000g) (Pharoah et al., 1990, 1996; Naulty et al., 1994; Hagberg et al., 1989, 1993, 1996; Topp et al., 1997; O'Shea et al., 1998). As low birth weight infants have a higher incidence of CP, this has resulted in an increasing prevalence of CP and changing patterns of CP, as well as a change in the survival rate of persons with CP (Evans et al., 1990; Hutton et al., 1994; Crichton et al., 1995).

Corresponding with improved neonatal care, there also has been marked improvement in the care of children with CP over the past 30 years (Evans et al., 1990). Children have access to improved health care, and new technologies allow them to have a better quality of life and
fewer complications. Meanwhile, social support systems have assisted parents in caring for children in their homes or with the assistance of associate or foster families. As a result, most children no longer live in institutions but live in their communities (Miller et al., 1978; Eyman et al., 1988; Bilsard et al., 1988).

Although the life expectancy of severely involved individuals is significantly less than that of the general population, more than 90% of children with CP live to adulthood (Evans et al., 1990). Even severely disabled children have about a 50% probability of surviving to 20 years of age (Hutton et al., 1994). However, there is very little information about the actual prevalence, survival rate, and associated issues of CP within BC. This information is important for the following reasons:

1. Parents want to know what to expect with regard to the survival, associated problems, and long term care of their children. For effective counseling, we must have more precise information on survival and these figures need to be relevant to conditions that exist in the community.

2. Health, education, social service and community planners need to have accurate information to ensure that services are available.

3. Increasingly, there are ethical questions being raised about quality of life issues, the aggressiveness of “rescue” efforts in the newborn period, and the rationing of services to certain segments of the population. These are critical issues and it is important to have accurate information about prevalence and survival when dealing with these issues.

Currently, the information on prevalence and survival is limited. Much reliance has been placed on the studies by Eyman and his colleagues (1990, 1993) who provided population-based information on survival of children and adults seen within the Department of Developmental Services in California. While important, this information cannot be easily extrapolated to
situations in BC and Canada because Eyman's studies, and others like them, do not provide sufficient information or because the criteria for inclusion or exclusion are not sufficiently well defined or are not consistent with the recording criteria used in BC. Although some preliminary work focusing on survival and influencing factors with type of CP, epilepsy and severity of mental retardation has been done in this area (Crichton et al., 1995), the findings are not sufficiently detailed to provide the information required.

6.2. Hypotheses and Rationale

In most developed industrial countries, the prevalence of CP has changed in the past four decades. The crude livebirth prevalence of CP from the 1950s through 1990 can be divided into two distinct trend eras: a period of decreasing prevalence during the 1950s and 1960s, and a period of increasing prevalence during the 1970s and 1980s. The first era is a period when perinatal care became better organized and good basic methods of treatment were established, whereas the second era is a period of development of mechanical ventilation and modern neonatal intensive care resulting in an increasing CP incidence attributable to improved survival of low birth weight infants (Hagberg et al., 1984, 1989, 1993; Pharoah et al., 1987, 1990, 1996; Takeshita et al., 1989; Stanley and Watson, 1988, 1992). However, a break in the successive increase in CP prevalence was reported in Hagberg's latest study (Hagberg et al., 1996) during the period 1987-1990. The prevalence of CP in low birth weight infants increased from the 1970s through the early 1980s in most countries except Norway (Meberg, 1990, 1995) and Slovenia (Kavcic and Perat, 1998); and the lower the birth weight group, the later the rise in prevalence of CP occurred. Meanwhile, the origin and type of CP also changed along with the changing trends in the prevalence of CP in different birth weight subgroups. There is, however, very little information about the actual prevalence within BC.
The evidence indicates that more than 90 percent of children will now survive well into their adult years (Evans et al., 1990). The survival rate of mildly and even moderately affected children approaches that of normal, unaffected children, at least for the first 20 years (Hutton et al., 1994), and the overall survival rate at 30 years is at least 87 percent (Crichton et al., 1995). However, the findings are not sufficiently detailed to provide sufficient information about prognostic factors such as maternal and perinatal conditions, etiology or origin of CP, motor and other functional disabilities, visual and hearing impairments, and respiratory, feeding and orthopaedic problems. The different trends observed and the different definitions and criteria used in the various studies make it difficult, if not impossible, to extrapolate the information so as to obtain specific insights about prevalence and survival rates of children with CP in BC.

Despite the lack of specific information, it is reasonable to hypothesize that:

1. The prevalence of persons with CP in BC has increased over the last 20 years.
2. There have been changes in the patterns of variables associated with prevalence, such as etiology, proportion of CP by birthweight, type of CP, and severity of CP.
3. Survival rate and prognostic factors of persons with CP in BC have changed.
4. Detailed information about the previous parts can be obtained by a retrospective study of BC medical charts.

6.3. Objectives

Given the lack of information about prevalence, survival rate and associated issues of CP in BC and given the inability to extrapolate information obtained in other areas to the situation within BC, we propose a retrospective study to extract detailed information on the prevalence and survival of persons with CP in BC; information detailed enough to enable BC health
professionals to make practical informed decisions when working with patients with CP and their families. To achieve this objective this study will develop and use a protocol that is accurate and precise enough to allow the information extracted to be usefully compared to other studies. Ideally this protocol could serve as the basis for a standardized protocol which would allow better comparison between future studies and thus greatly increase the value of such studies.

The objectives of this study are:

1. To estimate survival rate and related prognostic factors for patients with moderate or severe CP in BC.

2. To estimate the prevalence of moderate to severe CP in BC.

3. To try to ascertain whether or not a change in survival and prevalence trends is associated with different management in caring for persons with moderate or severe CP, e.g., early intervention, surgical therapy.

4. To identify any changes in the types and associated issues of CP associated with changes in prevalence.

6.4. Study Plan

6.4.1 Introduction — study overview

The proposed study will be a provincial retrospective study of prevalence and survival of persons with moderate to severe CP through two Health Services Linked Databases in BC. Data pertaining to prevalence and survival will be obtained from these two databases. Additional information relating to etiologic factors, functional abilities, associated problems, prognostic factors and management will be extracted from the medical charts of patients using the revised Cerebral Palsy Chart Review Protocol.
The extracted data will be stratified so as to assess time trends in the prevalence of CP and associated issues over the length of the study period. Finally, the extracted data will be subjected to standard statistical methods of primary and secondary analysis.

6.4.2. Definitions

6.4.2.1. Definition of CP

To be considered as a case, for the purposes of this study, a child must have met the following criteria (Pellegrino and Dormans, 1998):

1. The disorder is mainly in motor dysfunction, a significant problem with controlling movement and posture.
2. The disorders is a result of a disturbance or anomaly in early brain development.
3. The disorder is caused by nonprogressive insults or anomalies.

Diagnosis of CP is usually made within the first 3 years of life, but because this study also focuses on type and severity of CP, and because type and severity may take longer to ascertain, an upper age limit of 5 years is used.

6.4.2.2. Classification of CP

CP will be physiologically categorized as follows:

1. Spastic type (pyramidal, spastic extremities)
2. Dyskinesia (nonspastic, extrapyramidal, abnormal involuntary movement)
3. Ataxia (lack of balance and uncoordinated movement)
4. Atonia (lack of muscle tone)
5. Mixed type (spastic type with ataxia and/or dyskinesia)
6. Unclassified type
The *spastic subtypes* will be classified *topographically* depending on the number of limbs involved:

(1) **monoplegia** — involves one limb, usually is a variation of hemiplegia or diplegia.

(2) **hemiplegia** — lateralized one-half of the body affected.

(3) **diplegia** — both legs affected, slight upper limb involvement noted.

(4) **paraplegia** — involves the legs only, with normal upper limb function.

(5) **quadriplegia** — all limbs affected.

### 6.4.2.3 Etiology of CP

The etiology of CP will be recognized according to the time of occurrence of causative factors. These factors will be divided into prenatal, perinatal/neonatal, postneonatal, and unclassifiable factors. A case will be classified as “acquired” if the presumed insult to the brain occurs after the 28th day of life (postneonatal period). If there is uncertainty about the timing of the insult, the etiology will be presumed to be “idiopathic” or “congenital” (Pharoah et al., 1996). “Prenatal” will refer to the period from the first day of last menstruation to the onset of labor resulting in delivery; “perinatal” to the period from the onset of labor until the 7th day of life; “neonatal” to the period from birth to the first 28th day of life; and “postnatal” to the period from the 8th day of life onwards. “Prematurity” refers to birth that occurs before completion of the 37th week of gestation. Prematurity is further subdivided as follows: moderately preterm (32-36 weeks), very preterm (28-31 weeks), and extremely preterm (< 28 weeks). Low birth weight is a birth weight of < 2500g; very low birth weight is a birth weight of < 1500g; and extremely low birth weight is a birth weight of < 1000g.
6.4.2.4. Severity of CP

The severity of CP will be subdivided into five levels according to the criteria of Gross Motor Function Classification System (Russell et al., 1989; Russell et al., 1993; Palisano et al., 1997).

*Gross Motor Function* will be classified as follows:

(1) level I — walks without restrictions; limitations in more advanced gross motor skills.

(2) level II — walks without assistive devices; limitations walking outdoors and in the community.

(3) level III — walks with assistive mobility devices; limitations walking outdoors and in the community.

(4) level IV — self-mobility with limitations; children are transported or use power mobility outdoors and in the community.

(5) level V — self-mobility is severely limited even with the use of assistive technology.

Patients will be ranked as having mild, moderate or severe CP according to their level of ambulatory ability. Children on level one or two of the Gross Motor Function Classification System will be classified as mild cases. Children with level three ability will be classified as moderate. And children with level four or five ability will be classified as severe. Children who are too young or who died before they could be assessed will be recorded on the data chart as “unclassified.”

6.4.2.5. Associated dysfunctions of CP

*Mental ability* will be categorized according to the intelligence quotient (IQ) as follows:
(1) normal — IQ ≥ 85.

(2) mild mental retardation — IQ 70-84.

(3) moderate mental retardation — IQ 50-69.

(4) severe mental retardation — IQ < 50.

(5) the disability is thought to be severe to profound but is not testable because the subject lacks the cognitive skills to respond adequately to the testing.

If there has been more than one assessment, the most recent one will be used in the analysis of severity of mental dysfunction of CP, and the others will be used to study the progression of the child’s condition.

*Manual dexterity* will be classified as follows:

(1) normal — upper limbs unaffected.

(2) mild disability — some clumsiness of fine movements but able to feed and dress without assistance.

(3) moderate disability — able to feed and dress with difficulty.

(4) severe disability — unable to feed and dress without assistance.

(5) no functional use of hands.

*Visual impairment* will be classified as follows (Pharoah et al., 1998):

(1) mild — vision is > 20/60 in the better eye.

(2) moderate — vision is 20/60 to 20/200 in the better eye.

(3) severe — vision is < 20/200 in the better eye or there is no useful vision.

Accurate assessment of visual levels is often difficult in children with concomitant intellectual impairment.
Hearing loss is averaged across frequencies 0.5-4 kHz and the impairment is classified as follows (Pharoah et al., 1998):

1. Mild — hearing loss is 21-45 dB.
2. Moderate — hearing loss is 45-70 dB.
3. Severe — hearing loss is > 70 dB.

Assessment of accurate hearing levels is often difficult in children with concomitant intellectual impairment.

Epilepsy is defined as two or more afebrile seizures occurring after 28 days of life. Subjects with epilepsy will be classified according to the type and frequency of seizures as well as the need of medication for treatment. Epilepsy will be categorized as follows:

1. Generalized seizure.
2. Partial seizure.
3. Partial seizure with secondary generalization.
4. Infantile spasm.
5. Unclassified.

6.4.3. Sample Characteristics

For the evaluation of the prevalence and survival of children with CP, the proposed study cohort will consist of all cases of children with CP born between 1952 and 1993 and reported to the Health Surveillance Registry or the linked databases of the Centre for Health Services and Policy Research of BC or the Division of Vital Statistics with death certificates. All hospital chart data up to the end of 1998 of all ascertained cases will be reviewed. As this study will focus on survival and the prognostic factors of persons with moderate to severe CP,
the data retrieval through medical chart review will be performed only for moderate or severe cases. Because a confirmed diagnosis of CP and reliable physiological classification of CP is not always possible before the age of five (Stanley and Watson, 1992; Pharoah et al., 1996; Hagberg et al., 1996) this study will be limited to all children born by the end of 1993. The annual total number of livebirths in BC between 1952 and 1993 will be used as population denominator for evaluating the prevalence of CP.

6.4.3.1. Inclusion criteria

(1) This study will be limited to all persons with CP born by the end of 1993.

(2) The cohort for evaluating prevalence will include only those cases born in BC or elsewhere, between 1952 and 1993, whose mother was legally considered a resident of BC at the time of birth.

(3) The cohort for evaluating survival will include all cases, regardless of birthplace, resident in BC between 1952 and 1998.

(4) The study will focus only on cases classified as congenital or idiopathic CP by the end of 1998.

(5) Data retrieval through medical charts will be conducted only on moderate or severe cases as defined in section 6.4.2.4.

6.4.3.2. Exclusion criteria

(1) All cases with ambiguous diagnosis of CP who died before the age of 5 years will be excluded.

(2) Postneonatal cases of CP will be excluded.
6.4.3.3. Sample size estimate

In a previous study on life expectancy of persons with CP in BC, a total of 3189 cases were identified during the study period from 1952 through 1989 (Crichton et al., 1995). Although Crichton's definitions and criteria for ascertainment vary slightly from the ones proposed for this study, his numbers are still useful for obtaining a reasonable first approximation of the total number of cases during our study period. The total population in BC in 1989 was approximately 3.21 million and the total livebirths from 1952 through 1989 was approximately 1.43 million. Because the total population in BC at the end of 1993 increased to approximately 3.57 million and the total livebirths from 1952 through 1993 was approximately 1.61 million, an estimated 400 to 500 new cases will be identified from 1990 through 1993, bringing the estimated total number of cases to around 3700 during this proposed study period.

6.4.4. Conduct of the study

The validity of the study depends on a full ascertainment of cases, followed by extraction of relevant data. Ascertainment requires searching and evaluating all possible sources to locate all moderate and severe cases of CP reported in BC over the duration of the study period. Data is then extracted from all ascertained cases by reviewing chart records.

6.4.4.1. Ascertainment of cases

To ensure completeness of ascertainment of cases, the following sources of information will be used:

(1) Health Surveillance Registry (HSR).

This population-based registry serving the entire population of BC has been recording cases of birth defects, genetic disorders, and chronic disabilities since 1952.
Because the registry is maintained within the Division of Vital Statistics, it has access to such important sources as the physician’s notice of birth, hospital admission and separation forms for all children aged 7 years or younger in the province, death registrations and stillbirth records. New cases are added to the registry each year, and existing records are updated with new information. This ongoing registration and updating from many different sources greatly improves the accuracy and usefulness of the data base.

Data on individuals with disabilities are collected from more than 60 sources, including special schools and institutions, public health units, hospital discharge records of children under the age of seven years, and the agency of the provincial government which distributes allowances to disabled people from the age of 18 years (Baird, 1987). In addition, many community agencies, special clinics, lay organizations and physicians voluntarily register cases of handicapping disabilities with the registry.

Each case recorded in the computerized data base by the registry is given an identification number; and the name, full date of birth, sex, place of birth, parents’ names, mother’s maiden name, address, name of family physician and treatment facilities attended are recorded. A cross-reference is made to any other family members who are registered.

The diseases or disabilities are assigned on ICD-9 code number (World Health Organization, 1978), and the date of onset is recorded, if known. Age at registration is recorded, and an etiology code developed at the registry is assigned. The etiology codes include autosomal dominant, recessive, X-linked, chromosomal, multifactorial
and genetic, unspecified; there are also codes for various environmental causes (e.g., infection, trauma and nutrition) and for unknown cause.

(2) British Columbia Linked Health Database (BCLHD).

This is a provincial government funded database dealing with Health Policy Research. This linked health database collects the records from six health ministry files (medical services, hospital separations, drug descriptions, long term care, deaths, and births) and index them with individual’s personal health identification number (PHN) code. Linkage begins with files generated in the 1985/6 fiscal year and is current to the latest year. This database provides the following information: the basic characteristics of birth; date and cause of death; dates, length of stay, diagnosis, and procedure information from hospital encounters; and date and type of physician encounter, including diagnostic codes from physician billings database.

These diagnostic codes can be used for identification of cases associated with specified factors, issues and interventions. Most subjects who received specific surgical treatments such as gastrostomy, orthopaedic surgery, or neurosurgery were included in the database. Through this database, persons who have been registered with the Health Surveillance Registry are linked to the Medical Services Plan (MSP) to establish the patients’ MSP identities, thus giving access to all the patients’ available MSP data such as name of treating practitioner and hospital of treatment.

(3) Linkage between the Health Surveillance Registry database and the British Columbia Linked Health database.
Virtually all children with CP born in BC are registered in the Health Surveillance Registry, and many will have dual enrolment with the BCLHD. However, a very small minority of children with CP who migrated from other countries or patients associated with specific nutritional, orthopaedic or neuro-surgical problems were exclusively included in the BCLHD. Since the two databases are cross referenced this minority of cases can be quickly identified and extracted for survival analysis.

(4) *Death Registry*

A search will be made for all death certificates in which CP features as the underlying cause of death on Part I or as a significant condition contributing to the death on Part II of the certificate.

6.4.4.2. *Data extraction*

After ensuring the completeness of case ascertainment, data retrieval will proceed as follows:

1. Review charts for all ascertained cases to establish their severity.
2. To achieve more detailed data retrieval, the previously designed data extraction form, the revised Cerebral Palsy Chart Review Protocol, will be used.
3. Data retrieval will be performed by reviewing all cases of CP identified through the two databases and by extracting information from BC hospital chart records for the moderate or severe cases.
4. Records in which the diagnosis of CP was ambiguous will be reviewed by a pediatrician or pediatric neurologist. Children with ambiguous diagnostic information will be retained as presumptive cases, pending review of medical records.
by a physician. The final diagnosis of presumptive cases will be established through extensive review of Health Surveillance Registry records, hospital records, other medical records, and, if necessary, personal discussion with therapists or physicians familiar with the child.

(5) The latest data of utilization of Medical Services Plan will also display the name of the practitioner who treated the patient. If there is information lacking in specific cases the practitioner will be contacted directly to see if additional information can be supplied after consent has been obtained from the family.

(6) Information relating to specific associated problems and surgical interventions will be retrieved from the British Columbia Linked Health Database.

6.5. Analysis

6.5.1. Descriptive analysis

In order to assess the time trends in the prevalence of CP over the 42 year study period, subjects will be grouped into seven cohorts each with a 6 year period. This division into six year periods should give statistically meaningful sample sizes if the estimate of 3700 CP cases by the end of 1993 is correct. Since birth weight and gestational age are recognized as important factors in prevalence and severity of CP, the cohorts will be analyzed for the influence of these factors. According to birth weight each cohort will be analyzed for trends in proportion of CP by birth weight (the % of CP cases that fall within a given birth weight range), and trends in birth weight specific prevalence (the % of CP cases in a given birth weight range relative to the total number of livebirths in the province). According to birth weight and gestational age, respectively, each cohort will be analyzed for trends in etiological groups, trends in clinical type of CP, and trends in severity of CP. For birth weight, cases will be
divided into 4 groups: normal birth weight (≥ 2500g), low birth weight (<2500g), very low birth weight (< 1500g), and extremely low birth weight (<1000g). Similarly, according to gestational age, cases will be stratified as term (≥ 37 weeks), moderately preterm (32-36 weeks), very preterm (28-31 weeks), and extremely preterm (< 28 weeks).

6.5.2. Statistical analysis

To evaluate the prevalence and trends in CP subtypes, the chi-square (X²) test, the “trends in proportions” test, and the “significance test for comparing two proportions” will be used for statistical calculations (Armitage, 1983; Kirkwood, 1988). P values < 0.05 were considered statistically significant. A risk ratio measure with 95% confidence levels will be used to obtain a point estimate of the relative risk of mortality among subgroups of survivors to age of 5 years.

The Kaplan-Meier survival curve method (Kleinbaum, 1996) will be used with the statistical software packages SPSS 7.5 for Windows (1996) to estimate the length of survival and to display the data for the various classifications of the prognostic factors.

A Log-rank test will be used to assess differences in survival rates. The individual classification categories of a prognostic factor will also be compared globally using a Log-rank test to see if there are overall differences in survival rates. In order to examine the possible interactions of the prognostic factors on survival time, Cox’s proportional hazards model will be used (Kleinbaum, 1996). This method assumes that the survival function over time is the consequence of a baseline survival function which is modified by the effect of prognostic factors. The proportionality assumption of this model will be checked by reviewing the log (-log) plots of the Kaplan-Meier survival spots.
6.6. Ethics and Confidentiality

The Health Surveillance Registry and the Health Policy Registry databases both have a firm and clearly defined policy on personal identifying information. All personal identifying data are completely confidential and are accessible only to registry staff. Names and other information are available only to the agency that registered the case. In addition, the privacy of the registry data is protected through legislation in the form of an amendment to the Evident Act (Baird, 1987).

In this study the confidentiality requirement of the registries will be observed, and no personal identifying data will be singled out. The confidentiality of hospital or treatment centre records will also be respected.

6.7. Feasibility and Collaboration

BC, which has a population of approximately 4 million, is in a unique position to examine the prevalence and long term survival of children with CP. The Health Surveillance Registry has been in place since 1952 and provides records of children and adults with disabilities. This Registry, in combination with the information obtained from the database of the Centre for Health Services and Policy Research, provides the opportunity to examine a large, carefully documented population of children. As we mentioned above, through the Health Policy Research Centre persons who have been registered with the Health Surveillance Registry can be linked to the Medical Services Commission. The utilization of the Medical Services Plan can be linked to the names of these persons. A list of names of persons with CP can be obtained. The latest date of utilization of MSP will also display the name of the practitioner who treated the patient and the hospital where the patient visited.
After completing procedures for the ascertainment and allocation of cases, a formal proposal and application form for in-hospital research review will be sent to each hospital, and then data retrieval will be performed through reviewing the charts of each hospital.

6.8. Limitations

(1) Infants who died or migrated out of British Columbia in the first few months of life will be less likely to have been defined as “cases” in this study.

(2) Mild and possibly moderate cases who did not receive medical services under the diagnosis of CP or who did not register with the medical services databases as “cases” will be difficult to ascertain and classify.

(3) Children who were diagnosed as “cases” but who could not be followed up because they moved out of Canada or because their parents did not seek further medical assistance will not have a complete data record.

(4) Chart data for children adopted from other countries, or for children who migrated from other countries, may be limited, especially as regards maternal pregnancy and delivery records and neonatal records.

(5) Because persons who move into the province may have received different standards of health care and because the move itself may have caused stress and adjustment problems which could affect their life expectancy their inclusion may slightly skew survival results.

(6) As the study period goes back to 1952, some medical charts may no longer be available, thus preventing complete chart retrieval.
6.9. Significance of this proposed study

While CP is costly (Waitzman et al., 1995), the overall prevalence rate and the survival rate of individuals with CP have increased in the last thirty years. At present, there is no actual information about the prevalence of CP and the associated issues in BC. The information about survival and prognostic factors in previous studies is not sufficiently detailed.

This proposed study will provide more detailed information about prevalence, etiology, type, functional ability, associated problems, and survival rate of patients with CP in BC. This information will be important for health, education, social service and community planners to ensure that services will be available. The information will also be important for clinicians and families to improve decision making around issues of care. Furthermore, this large and long term study will extract information relating etiologies or causative factors and will be helpful in assessing methods and techniques for the prevention of CP.
Chapter 7. Overall Summary and Discussion

During the last four decades the prevalence of cerebral palsy (CP) has changed in most industrialized countries. Improvements in obstetrical and neonatal care have dramatically reduced the perinatal mortality rate among low birth weight infants. This change in mortality has resulted in an increasing prevalence of CP and in changing patterns of CP, as well as influencing the survival rate of CP. Corresponding with improved neonatal care, there also has been marked improvement in the care of children with CP over the last 30 years. The increase in prevalence and life expectancy of persons with CP naturally has important implications for health, education and social services. However, at present, there is very limited information about the prevalence of CP and its associated issues in British Columbia (BC). While some preliminary work has been done regarding the survival of persons with CP, the information is not sufficiently detailed to be of value to future planning.

This thesis started with a general review of CP. Among the aspects examined were historical background, definition, epidemiology, etiology, classification, associated conditions, pathological findings, diagnosis, treatment, prevention, and outcome of CP. The thesis then went on to review current population based studies for changing trends in the prevalence of CP and for survival of children with CP. Drawing upon these studies a data extraction form — the Cerebral Palsy Chart Review Protocol — was then developed and a pilot study was conducted to assess the feasibility of this protocol, along with a proposal for defining the prevalence and survival of children with CP in British Columbia.

To evaluate the changing trends in prevalence and associated issues of CP in the past decades, several computerized bibliographic databases from 1966 to 1998 were searched for population-based articles written in English and relating to CP prevalence time trends.
Although all the studies evaluated were published after 1975, some of them contained data going back as far as 1954. Concurrent with a decrease in perinatal mortality rates, the proportion of low birthweight infants with CP increased from 10-35% in the 1960s to 35-55% in the 1980s, with extremely low birthweight cases (<1000 gm) making a sizeable contribution to the total in the late 1980s and early 1990s. The crude prevalence of CP declined from the 1950s through the 1960s, and then increased progressively in the 1970s and the early 1980s in most countries except in Norway and Australia. In the late 1980s, there appears to have been a break in the trend of increasing prevalence in Sweden and in Slovenia. The prevalence of low birthweight infants with CP increased in the 1970s and 1980s in most studies while the prevalence declined from 1981 to 1990 in Slovenia. In Sweden, the degree of disability among preterms also decreased in the late 1980s. In analyzing the origins of CP, perinatal origin was predominant among children with CP born preterm; whereas prenatal origin was predominant among children with CP born at term. The most frequent criterion for a prenatal etiology, based on CT/MRI findings, was periventricular leukomalacia or atrophy; whereas the most frequent criterion for a peri/neonatal etiology was intracerebral hemorrhage/stroke. Alternatively, perinatal or intrapartal asphyxia was seldom the cause of CP, (3 to 13 % of cases). Spastic diplegic CP characterized the large majority of preterm cases, whereas intrapartum events were closely related to the pathogenesis of hemiplegia. Recent results suggest that improvements in perinatal care can reduce the incidence and severity of perinatal damage. More investigation of the causative factors of CP and continuing observation of the changes in the prevalence of CP should provide new clues about etiology and perhaps help in the treatment and prevention of CP.

The survival rate and prognostic factors for CP were also evaluated through a search of journal articles in several computerized bibliographic databases from 1966 to 1998. Only five
population-based articles focusing on survival of persons with CP were found. The number of subjects in each study was 64, 732, 1251, 3187 and 12719 respectively. Because of its small sample size and the limitations of statistical analysis, the 64 subject article was not included in this paper. The study duration ranged from 10 to 38 years with most individuals in the studies followed for at least 5 years. The scientific quality of the selected articles was assessed for applicability and validity. The data in these studies show that the factors influencing survival of CP include mobility, severe mental retardation, manual dexterity, feeding skills, epilepsy, birth weight, and gestational age. Some other factors, such as orthopaedic, visual and hearing impairments, feeding problems, respiratory infection, and technical advances and improvements in the management of these problems, might influence length of survival and quality of life as well. The survival rate of mildly and even moderately affected children approaches that of normal, unaffected children, at least for the first 20 years. Even children with severe disability have about a 50% probability of surviving to 20 years of age. The overall survival rate at 30 years is at least 87 percent. This high survival rate should prompt a reappraisal of the long term needs of people with CP, as well as of their medical care plans.

Although the results of these studies contribute important and valuable information about the prevalence and survival of persons with CP, there are methodological problems in the current literature. One of the difficulties in evaluating the papers is the lack of a standardized protocol. Researchers working in different countries, sometimes even within the same country, tend to use different definitions of CP, different criteria of inclusion and ascertainment, different associated issues in the prevalence of CP, and different prognostic factors for evaluating the survival of persons with CP. What is needed is a standardized protocol with very precise definitions, clear criteria of inclusion, consistent ascertainment, and as complete consideration of influencing factors and variations as possible.
Because of the limitation of the current literature, a proposal for defining the prevalence and survival of children with CP in BC was designed. To perform this study, the first step was to develop a Cerebral Palsy Chart Review Protocol to extract specific information relating to the prevalence and survival of children with CP from hospital or treatment-centre based records in BC. To assess the feasibility of the protocol a pre-test was completed on a convenience sample of charts from a British Columbia hospital for children with disabilities. The pre-test study results led to some revisions of the protocol: sections on visual and hearing impairments were added, the neonatal record was simplified, the sections on feeding and orthopaedic problems were modified, and mobility and manual dexterity were added as criteria to the section on level of disability. The new Chart Review Protocol permits the severity of CP to be evaluated according to ambulation skills, mental ability, manual dexterity, and visual and hearing impairments. The revised protocol also allows for easy extraction of information relating to etiological factors, functional disabilities and associated problems. The pilot study proved the form’s value as a valid extraction method. The form’s only apparent drawback is its exhaustive nature, as the patient records must be analyzed carefully to ensure that all desired pieces of information are extracted.

For the proposed study, we hypothesize that the prevalence of children with CP in BC has increased and the patterns of associated variables have changed over the last 20 years, and the survival rate and the prognostic factors of children with CP have changed. The study will be a retrospective study of prevalence and survival of children with CP through the Health Services Linked Databases of the Health Surveillance Registry and the Health Policy Research Centre linked to the Medical Services Commission in BC. Data pertaining to prevalence and survival will be obtained from these two databases; however, additional information relating to etiologic factors, functional abilities, and associated problems and management will be extracted from the
medical charts of patients using the revised Cerebral Palsy Chart Review Protocol. The proposed study will perform case ascertainment, data extraction, and stratification of cases and statistical analysis using the definitions and eligibility criteria developed in this thesis.

Some limitations may be encountered in this proposed provincial retrospective study. Infants who died in the first few months of life will be less likely to have been defined as "cases". Minimal or mild cases who did not receive medical services under the diagnosis of CP will be difficult to ascertain. Chart data of children with CP may be insufficient due to failure to follow up or due to migrating out of Canada. Also, maternal and neonatal records may be limited for children put into foster care or for children who had migrated from foreign countries. There may also be some confusion or gaps in information surrounding patients who have sought out treatment from several different doctors or hospitals or who have even rejected conventional medicine for alternative therapies.

In addition, the study must address several methodological issues. First, paediatricians, neurologists and orthopaedic surgeons have been trained differently, and not all agree on the exact motor diagnosis and may use different classification systems in the cases reviewed. To ensure consistency we have defined terms relating to definition, classification, etiology, severity, and eligibility criteria for CP. Second, to ensure completeness of ascertainment of cases, several sources of information will be used, including two Health Services databases and all death certificates. Third, records in which the diagnosis of CP was ambiguous will be reviewed by a paediatrician or paediatric neurologist. The final diagnosis of presumptive cases will be established through extensive review of Health Surveillance Registry records, hospital records, other medical records, and, if necessary, personal discussion with therapists or physicians familial with the child. Fourth, to ensure as complete a extraction of the data as possible, the Medical Services Databases will be checked to make sure all hospital or treatment-centre
records have been searched. Finally, the subjects will be stratified according to birth weight, gestational age, and study period to evaluate the prevalence trends and role of confounding; and statistical survival analysis will be used to assess the survival rate and possible connections between different prognostic factors.

Generally, this proposed study should provide important information for families and clinicians and will improve decision making around issues of care. The information will also be useful for more effective planning of health, education and social services. Furthermore, information relating to causative factors and associated issues of CP will be valuable in assessing methods and techniques for the prevention of CP.
Bibliography


Mutch L, Alberman E, Hagberg B et al. (1992) Cerebral palsy epidemiology: where we are now and where are we going? Dev Med Child Neurol 34: 547-55.


Appendix A: Cerebral Palsy Chart Review Protocol

Summary Data
Case #: ____________________
Chart #: ____________________
Birth Date (D/M/Y): __/__/___
Sex: ___ M ___ F
Birth Hospital #: ______________
MSP #: ______________________
Registration Date:__/__/___
Age: ___ Y ___ M

1. Classification of Cerebral Palsy

<table>
<thead>
<tr>
<th>ICD-9-CM Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>__ spastic type: ___ monoplegia ___ hemiplegia ___ paraplegia ___ diplegia</td>
</tr>
<tr>
<td>__ quadriplegia ___ other</td>
</tr>
<tr>
<td>__ dyskinetic type: ___ athetosis ___ chorea ___ dystonia ___ tremor</td>
</tr>
<tr>
<td>__ rigidity ___ other</td>
</tr>
<tr>
<td>__ ataxic type: ___ congenital cerebellar ataxia ___ other</td>
</tr>
<tr>
<td>__ atonic type: ___ diplegia ___ congenital (simple) ___ other</td>
</tr>
<tr>
<td>__ mixed type --- predominant type: ____________ distribution: ____________</td>
</tr>
<tr>
<td>__ unclassified</td>
</tr>
<tr>
<td>__ MI (missing information or incomplete data) ___ other</td>
</tr>
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2. Etiology of CP

<table>
<thead>
<tr>
<th>ICD-9-CM Code</th>
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<tbody>
<tr>
<td>__ Prenatal factors</td>
</tr>
<tr>
<td>__ genetic factors ___ Family History ___ chromosomal abnormality</td>
</tr>
<tr>
<td>__ acquired disease during gestation ___ abnormal fetal position</td>
</tr>
<tr>
<td>__ verified congenital infection ___ other</td>
</tr>
<tr>
<td>__ Perinatal/neonatal factors</td>
</tr>
<tr>
<td>__ hypoxic ischemic encephalopathy ___ intracerebral hemorrhage</td>
</tr>
<tr>
<td>__ periventricular hemorrhage ___ sepsis or CNS infection</td>
</tr>
<tr>
<td>__ confirmed brain edema or evidence of neonatal shock ___ other</td>
</tr>
<tr>
<td>__ Postneonatal factors</td>
</tr>
<tr>
<td>__ traumatic injuries ___ cerebral anoxia ___ toxic factors ___ seizure</td>
</tr>
<tr>
<td>__ vascular accidents ___ infections ___ brain tumors ___ other</td>
</tr>
<tr>
<td>__ Prematurity</td>
</tr>
<tr>
<td>__ low birth weight ___ very low birth weight ___ extreme low birth weight, with:</td>
</tr>
<tr>
<td>__ frequent prolonged apnea ___ periventricular hemorrhage</td>
</tr>
<tr>
<td>__ intracerebral hemorrhage ___ chronic lung disease ___ other</td>
</tr>
<tr>
<td>__ Unclassified or __ Idiopathic ___ MI</td>
</tr>
</tbody>
</table>

3. Severity of CP (according to Gross Motor Function Classification System)

<table>
<thead>
<tr>
<th>level 1</th>
<th>level 2</th>
<th>level 3</th>
<th>level 4</th>
<th>level 5</th>
<th>MI</th>
</tr>
</thead>
</table>

4. Mental retardation

<table>
<thead>
<tr>
<th>ICD-9-CM Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>__ normal ___ mild ___ moderate ___ severe or profound</td>
</tr>
<tr>
<td>__ suspected severe or profound but not testable ___ other ___ MI</td>
</tr>
</tbody>
</table>
5. Epilepsy

ICD-9-CM Code of etiology

- non-existent
- generalized
- partial
- infantile spasm
- partial with secondary generalization
- unclassified
- other

6. Feeding problems

ICD-9-CM Code

- N-G tube feeding
- TPN
- G-E reflux
- gastrostomy
- fundoplication
- jejunostomy
- other

7. Respiratory problems

ICD-9-CM Code

- frequent respiratory infection
- documented aspiration pneumonia
- need respiratory equipment
- supportive treatment
- documented reactive respiratory airway disease (e.g. asthma)
- other

8. Orthopaedic problems and management

ICD-9-CM Code

- scoliosis
- kyphosis
- lordosis
- dystonia
- other

- support treatment:
- operation:
- other treatment:

9. Others

ICD-9-CM Code

- infectious problems:
- any other major medical condition:

10. Management

ICD-9-CM Code

- early intervention
- medicine
- rehabilitation
- surgical procedure
- other

11. Survival status

ICD-9-CM Code

- alive
- resolutive
- MI
- deceased

date of death (D/M/Y): / / 

time (Hr: Min): :

age of death (M/Y): / 

cause of death:

Cerebral palsy Chart Review Protocol

I. General Data

Case #: ______
Chart #: ______
Birth Date (D/M/Y): / / /
Sex: ___M ___F

Birth Hospital #: ______
MSP #: ______
Registration Data: / / /
Age: ___Y___M

Maternal Data
Birth Hospital #: ______
Infant Chart #: ______
Birth Date (D/M/Y): / / /
Admission Date (D/M/Y): / / /
Marital State: ______
Social Service: ___ Yes ___ No

Maternal Chart #: ______
MSP #: ______
Age: _____ Yrs
Separate Date: / / /
Ethnicity: ______

Paternal Data
Age: ______ Yrs

Ethnicity: ______

Family History

II. Maternal Record

Pregnancy complications (conditions originating in the prenatal period)
Major Complication : ___ Yes ___ No ___ MI (missing information)
If yes, highlight and summarize: i.e. gestational age wk. (when first noted), treatment (when started), treatment type (none, medication, bed rest, hospitalized) and values of measurements or Lab results (where applicable).

Complications of labour and delivery (complications originating at onset of or during labour and delivery) : ___ Yes ___ No ___ MI
If yes, highlight and summarize stage of labour, onset time and duration of complication (if applicable).

Fetal factors (abnormal conditions of the fetus known prior to delivery)
___ Yes ___ No ___ MI
If yes, record gestational age when diagnosed, treatment (if any), treatment type and value of measurement and Lab results.

III. Newborn record: unless otherwise specified, the attributes may occur at any time prior to neonate’s discharge home (or death). If an attribute is not mentioned in the chart, assume that it was either normal, not present or in the normal range.

Chart #: Date of birth (D/M/Y): / / 
Time of birth (hr./min): ____ Sex: ___ M ___ F  
___ single ___ multiple: ___ 1st born ___ 2nd born  
gestation (by date first choice): ___ wk. by exam: ___ wk. 
birth weight: ___ gm length: ___ cm head girth: ___ cm

Condition at birth
APGAR score: 1 min ___ 5min ___ 10min ___ 15min ___ 20min ___  
Respiration (< 20 min of life): ___ normal ___ abnormal ___ spontaneous
___ none for ___ min ___ regular ___ irregular ___ other ___ MI

Resuscitation: record any / all occurrences prior to discharge home (or death)
___ Yes ___ No ___ MI
If yes, highlight and record infants age at onset (min) and length of time to establish normal respiration < 20 min of life. If resuscitation is initiated at 20 > min of life, summarized under respiratory distress.
___ oxygen: maximal O2 level ___ length of time (hr/min) ___/
___ mask ___ intubation ___ both
___ bag ___ IPPV ___ both
length of time to first spontaneous respiration ___ min
___ tracheal aspiration ___ cardiac massage ___ epinephrine
Meconium: ___ Yes ___ No ___ onset time (hr/min) ___ / ___ thick
___ present in amniotic fluid ___ staining on skin or cord
___ present below the vocal cords

Summary:

Delivery trauma (infant trauma resulting from labour/delivery)
___ Yes ___ No ___ MI  
Time of birth (hr/ min): ___:
If yes, highlight and describe type of injury, age when noted, consults, test.

Neonatal Feeding problem:
___ No ___ abnormal or difficulties: length of time ___ days ___ MI
mode: ___ breast ___ bottle ___ N/G ___ TPN

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**Neonatal Respiratory disorders**
Respiratory conditions continuing beyond initial resuscitation or those that developed after the first 20 minutes of life:  
- Yes  
- No  
- MI  
If yes, highlight and summarize age when noted, consults, test results.

**Neonatal non-neurological changes** (other abnormal conditions not previously recorded)  
- Yes  
- No  
- MI  
If yes, highlight and summarize age when noted, consults, test results.

**Neonatal neurological condition**: abnormal neurological conditions present at birth or those that anytime prior to discharge:  
- Yes  
- No  
- MI  
If yes, highlight and record onset time, duration and abnormal summary if neurologist consulted, type of injury, age when noted, consults, tests.

<table>
<thead>
<tr>
<th>Tension fontanel:</th>
<th>normal</th>
<th>abnormal:</th>
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</thead>
<tbody>
<tr>
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<td>normal</td>
<td>abnormal:</td>
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<tr>
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<td>abnormal:</td>
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<tr>
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<td>abnormal:</td>
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<tr>
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<tr>
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<td>hypertonia / tight</td>
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<tr>
<td>onset</td>
<td>duration (day/hr)</td>
<td>(if identified)</td>
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<tr>
<td>present</td>
<td>&lt; 2 hr.</td>
<td>2-120 hr.</td>
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<tr>
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<td>obtundation</td>
<td>coma</td>
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<tr>
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<tr>
<td>Cranial nerve function:</td>
<td>normal</td>
<td>abnormal:</td>
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<tr>
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<tr>
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<tr>
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<td>abnormal: (describe)</td>
<td></td>
</tr>
</tbody>
</table>

*Present < 2 hr.*

*2-120 hr.*

*> 120 hr.*
normal abnormal: (describe)

MRI: date (D/M/Y) ___/___/___ age: ___/___ (day/hr.)

normal abnormal: (describe)

other:

Neonatal Jitteriness / seizure: ___ Yes ___ None ___ MI
  jittery / tremor: onset time post birth (day/hr) ___/___ duration ___
  seizure: onset time post birth (day/hr) ___/___ duration ___

Drugs used for treatment of seizure:

Summary:

IV. Cerebral Palsy (C.P.)

age: ___ Y ___ M (at time of most recent functional estimation)
age of diagnosis of C. P.: ___ Y ___ M
social service: ___ Yes ___ No If yes, name of service institution: ____________

4.1. Classification of Cerebral Palsy

___ spastic type: ___ monoplegia ___ hemiplegia ___ paraplegia ___ diplegia
     ___ quadriplegia ___ other
___ dyskinetic type: ___ athetosis ___ chorea ___ dystonia ___ tremor
     ___ rigidity ___ other
___ ataxic type: ___ congenital cerebellar ataxia ___ other
___ atonic type: ___ diplegia ___ congenital (simple) ___ other
___ mixed type — predominant type: ____________ distribution: ____________
     ___ other ___ MI

4.2. Etiology of C.P.

___ prenatal factors

___ genetic factors: chromosomal finding __________
___ Family history: __________
___ acquired during gestation:
     ___ confirmed maternal infection ___ irradiation
     ___ prenatal anoxia: ___ placental abnormalities or ___ maternal anoxia
     ___ hemorrhage during pregnancy ___ maternal toxemia
     ___ fetal cerebral hemorrhage
     ___ predisposition to miscarriage
     ___ metabolic disturbances during pregnancy
     ___ other
**Perinatal/Neonatal Factors**

- Hypoxic Ischemic Encephalopathy:
  - APGAR score: 5 min __ 10 min __ 20 min __
  - Mechanical respiratory obstruction e.g. coiling of cord
  - Meconium aspiration __ CPD __
  - Prolonged fetal bradycardia __ other __
  - Intracerebral hemorrhage
  - Mechanical factors depending upon the type of delivery
    - Prolong labor __ CPD __ other __
  - Periventricular hemorrhage: Grade __
  - Confirmed brain edema or evidence of neonatal shock __
  - Sepsis or CNS infection __ other __

**Postneonatal Factors**

- Traumatic injuries __ Infections __ Toxic factors __ Seizure __
- Vascular accident __ Cerebral anoxia __ Brain tumors __ other __

**Prematurity**

- Low birth weight (1501-2300), or very low birth weight (<1501), with:
  - Frequent prolonged apnea __ Periventricular hemorrhage: grade __
  - Intracerebral hemorrhage __ Chronic lung disease __ other __
  - Unclassified or Idiopathic __ other __ MI __

**4.3. Severity of C.P. (according to Gross Motor Function Classification System)**

- Level 1 __ Level 2 __ Level 3 __ Level 4 __ Level 5 __ MI __

**4.4. Level of disability**

**4.4.1 Motor domain**

- Rolling and sitting
  - 1 = Does not lift head when lying on stomach
  - 2 = Lifts head when lying on stomach
  - 3 = Lifts head and chest using arm support when lying on stomach
  - 4 = Rolls from side to side
  - 5 = Rolls from front to back only
  - 6 = Rolls from front to back and back to front
  - 7 = Maintains sitting position with minimal support for at least five (5) minutes
  - 8 = Sits without support for at least five (5) minutes
  - 9 = Assumes and maintains sitting position independently

- Hand use
  - 1 = No functional use of hand
  - 2 = Uses raking motion or grasps with hand
  - 3 = Uses thumb and fingers of hand in opposition
  - 4 = Uses fingers independently of each other
arm use
1= no functional use of arm
2= moves arm from shoulder but does not extend or flex arm (i.e., does not have control of elbow joint)
3= partially extends arm
4= fully extends arm
crawling and standing
1= does not crawl, creep or scoot
2= crawls, creep, or scoots
3= pulls to a standing position
4= stands with support for at least one (1) minute
5= stands unsteadily alone for at least one (1) minute
6= stands well alone, balances well for at least five (5) minutes
ambulation
1= does not walk
2= walks with support
3= walks unsteadily alone at least ten (10) feet
4= walks well alone at least twenty (20) feet, balances well
climbing stairs
(rate uses of ramps for persons using wheelchairs)
N= no opportunity to use stairs (or ramps)
1= does not move up or down stirs (or ramps)
2= moves up and down stairs (or ramp) with help
3= moves up and down stairs (or ramps) with hand rail independently
4= moves up and down stairs (or ramps) without need for handrail
wheelchair mobility
N= does not use wheelchair
1= sits in wheelchair, does not move wheelchair by self
2= assists in moving wheelchair
3= moves self with some bumping and/or difficulty in steering
4= moves or guides chair independently and smoothly

4.4.2. Other functional domain
eating skills: ___ finger feeding ___ unable to feed by self ___ tubal feeding ___ other
___ MI
toilet: ___ trained ___ partial trained ___ not trained ___ other ___ MI
receptive nonverbal: ___ communication ___ some ___ none ___ other ___ MI
speech skills: ___ well expression ___ poor expression ___ can not speech ___ other ___ MI
recognize language ability: ___ well ___ moderate ___ poor ___ other ___ MI
social response: ___ well ___ moderate ___ poor ___ other ___ MI
4.5. **Mental retardation in C.P.**

- mild
- moderate
- severe or profound

IQ: ______  age of test (Y/M) _____/_____  method: ______

- suspected severe to profound but not testable  ______ other  ______ MI

4.6. **Epilepsy in C.P.**

- non-existent
- existent: onset time (Y/M) _____/_____  ___ MI

- frequency of seizure: < 1 per day  ___  ≥ 1 per day  ___ other:

- type of seizure: ___ generalized  ___ partial  ___ infantile spasm

- partial with secondary generalization  ___ unclassified

- other  ___ MI

- treatment of seizure:

- outcome of epilepsy after treatment:

- seizure free off medication  ___  seizure free on medication

- occasionally seizure on medication  ___  frequently seizure on medication

- other  ___ MI

4.7. **Feeding problems in C.P.**

- onset time of feeding problem (Y/M) _____/_____  

- treatment of feeding problems:

- N- G tube feeding  ___  TPN  ___ other  ___ MI

- operative treatment: ___ gastrostomy  ___ fundoplication

- jejunostomy  ___ other

- when:

- where:

- why:

4.8. **Respiratory problems in C.P.**

- frequent respiratory infection  ___  documented aspiration pneumonia

- need respiratory equipment supportive treatment

- documented reactive respiratory disease (e.g.: asthma)

- other  ___ MI

4.9. **Orthopaedic problems in C.P.**

- scoliosis
- kyphosis
- dystonia
- other

- managed with:

- medicine: ___ Baclofen for spasticity  ___ other

- supportive treatment: ___ standing frame  ___ posterior walker

- electric wheelchair  ___ night splints to ankles

- soft splints  ___ ankle-foot orthoses

- footwear attached with/without below-knee calipers

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4.10. Management of C. P.

the time of diagnosis as C.P. (D/M/Y): ___/___/___
age of start of treatment (Y/M): ___/___
location of care: ___ at home ___ institution ___ hospital ___ foster family
___ alternative home care ___ other ___ MI
type of treatment: ___ early intervention
___ medicine
___ rehabilitation
___ surgical procedure
type of operation:
___ other ___ MI

4.11. Outcome:
___ alive ___ deceased ___ resolutive ___ MI
prominent problems:
___ respiratory problem
___ infectious problem
___ feeding problem
___ epilepsy
___ orthopaedic problem
___ other
date of death (D/M/Y): ___/___/___
time (Hr : Min): ___:
age of death (M/Y): ___/
cause of death:

4.12. Summary:
Appendix B: Revised Cerebral Palsy Chart Review Protocol

Summary Data

<table>
<thead>
<tr>
<th>Case #:</th>
<th>Birth Hospital #:</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chart #:</td>
<td>MSP #:</td>
</tr>
<tr>
<td>Birth Date (D/M/Y):</td>
<td>Registration Date:</td>
</tr>
<tr>
<td>Sex: M __ F</td>
<td>Age: ___ Y ___ M</td>
</tr>
</tbody>
</table>

1. Classification of Cerebral Palsy (CP)

<table>
<thead>
<tr>
<th>ICD-9-CM Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. spastic type: ___ monoplegia ___ hemiplegia ___ paraplegia</td>
</tr>
<tr>
<td>___ diplegia ___ quadriplegia ___ other</td>
</tr>
<tr>
<td>2. dyskinetic type: ___ athetosis ___ chorea ___ dystonia ___ tremor</td>
</tr>
<tr>
<td>___ rigidity ___ other</td>
</tr>
<tr>
<td>3. ataxic type: ___ congenital cerebellar ataxia ___ other</td>
</tr>
<tr>
<td>4. atonic type: ___ diplegia ___ congenital (simple) ___ other</td>
</tr>
<tr>
<td>5. mixed type ___ predominant type: ___ distribution: ___ other</td>
</tr>
</tbody>
</table>

2. Etiology of CP

<table>
<thead>
<tr>
<th>ICD-9-CM Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Prenatal factors</td>
</tr>
<tr>
<td>___ genetic factors ___ family history ___ chromosomal abnormality</td>
</tr>
<tr>
<td>___ acquired disease during gestation ___ abnormal fetal position</td>
</tr>
<tr>
<td>___ verified congenital infection ___ other</td>
</tr>
<tr>
<td>2. Perinatal/neonatal factors</td>
</tr>
<tr>
<td>___ hypoxic ischemic encephalopathy ___ intracerebral hemorrhage</td>
</tr>
<tr>
<td>___ periventricular hemorrhage: grade ___ sepsis or CNS infection</td>
</tr>
<tr>
<td>___ confirmed brain edema or evidence of neonatal shock ___ other</td>
</tr>
<tr>
<td>3. Postneonatal factors</td>
</tr>
<tr>
<td>___ traumatic injuries ___ cerebral anoxia ___ toxic factors</td>
</tr>
<tr>
<td>___ seizure ___ vascular accidents ___ infections ___ brain tumors</td>
</tr>
<tr>
<td>___ other</td>
</tr>
<tr>
<td>4. Prematurity</td>
</tr>
<tr>
<td>___ low birth weight ___ very low birth weight</td>
</tr>
<tr>
<td>___ extreme low birth weight, with:</td>
</tr>
<tr>
<td>___ frequent prolonged apnea ___ periventricular hemorrhage</td>
</tr>
<tr>
<td>___ intracerebral hemorrhage ___ chronic lung disease ___ other</td>
</tr>
<tr>
<td>5. Unclassified ___ Idiopathic ___ MI</td>
</tr>
</tbody>
</table>

3. Severity of CP (according to Gross Motor Function Classification System)

<table>
<thead>
<tr>
<th>ICD-9-CM Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>___ level 1 ___ level 2 ___ level 3 ___ level 4 ___ level 5 ___ MI</td>
</tr>
</tbody>
</table>

4. Mental retardation

<table>
<thead>
<tr>
<th>ICD-9-CM Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>___ normal ___ mild ___ moderate ___ severe or profound</td>
</tr>
</tbody>
</table>
5. Manual dexterity  
*ICD-9-CM Code* 
- normal 
- mild disability 
- moderate disability 
- severe disability  

6. Epilepsy  
*ICD-9-CM Code of etiology* 
- non-existent 
- generalized 
- partial 
- infantile spasm 
- partial with secondary generalization 
- unclassified 
- other  

7. Feeding problems  
*ICD-9-CM Code* 
- N-G tube feeding 
- TPN 
- G-E reflux 
- gastrostomy 
- fundoplication 
- jejunostomy 
- other  

8. Respiratory problems  
*ICD-9-CM Code* 
- frequent respiratory infection 
- documented aspiration pneumonia 
- need respiratory equipment supportive treatment 
- documented reactive respiratory airway disease (e.g. asthma) 
- other  

9. Orthopaedic problems and management  
*ICD-9-CM Code* 
- scoliosis 
- kyphosis 
- lordosis 
- dystonia 
- other 
- support treatment: 
- operation: 
- other treatment:  

10. Visual impairment  
*ICD-9-CM Code* 
- Cortical visual impairment 
- complete blindness 
- field defect 
- strabismus 
- amblyopia 
- nystagmus 
- other 
- visual dysfunction: 
- mild 
- moderate 
- severe  

11. Hearing Impairment  
*ICD-9-CM Code* 
- complete deafness 
- partial deafness 
- neurosensory deafness 
- other 
- hearing loss: 
- mild 
- moderate 
- severe  

12. Others  
*ICD-9-CM Code* 
- infectious problems: 
- any other major medical condition:  

13. Age of confirmed diagnosis  
*Y* *M*  

14. Management  
*ICD-9-CM Code* 
- early intervention 
- medicine 
- rehabilitation 
- surgical procedure 
- other  

15. Outcome  
- alive 
- dead  

date of death (D/M/Y):  
- time (Hr:Min):  
- age of death (M/Y):  
- cause of death:  
*ICD-9-CM Code*
Cerebral Palsy Chart Review Protocol

I. General Data

Case #: 
Chart #: 
Birth Date (D/M/Y): / / 
Sex: M F

Maternal Data
Birth Hospital #: 
Infant Chart #: 
Birth Date (D/M/Y): / / 
Admission Date (D/M/Y): / / 
Marital State: 
Social Service: Yes No

Paternal Data
Age: Yrs

Family History

II. Maternal Record:

Pregnancy complications (conditions originating in the prenatal period)
Major Complication: Yes No MI (missing information)
If yes, highlight and summarize: i.e. gestational age wk. (when first noted),
treatment (when started), treatment type (none, medication, bed rest, hospitalized)
and values of measurements or Lab results (where applicable).

Complications of labour and delivery (complications originating at onset of or
during labour and delivery): Yes No MI
If yes, highlight and summarize stage of labour, onset time and duration of
complication (if applicable).

Fetal factors (abnormal conditions of the fetus known prior to delivery)
Yes No MI
If yes, record gestational age when diagnosed, treatment (if any), treatment type
and value of measurement and Lab results.
III. Newborn record: unless otherwise specified, the attributes may occur at any time prior to neonate’s discharge home (or death). If an attribute is not mentioned in the chart, assume that it was either normal, not present or in the normal range.

Chart #: __________ Date of birth (D/M/Y): __/__/__
Time of birth (hr./ min): __:__ Sex: ___ M ___ F
___ single ___ multiple: ___ 1st born ___ 2nd born
gestation (by date first choice): ___ wk. by exam: ___ wk.
birth weight: ___ gm length: ___ cm head girth: ___ cm

**Resuscitation:** record any/all occurrences prior to discharge home (or death)
___ Yes ___ No ___ MI
If yes, highlight and summarize: (eg. time and method of resuscitation, pH of umbilical blood, method of resuscitation)
length of time to first spontaneous respiration ___ min
___ tracheal aspiration ___ cardiac massage ___ epinephrine
Meconium: ___ Yes ___ No onset time (hr/min) ___ / ___
___ thick ___ present in amniotic fluid
___ staining on skin or cord ___ present below the vocal cords

**Delivery trauma** (infant trauma resulting from labour/delivery)
___ Yes ___ No ___ MI
presentation of labour: __________
If yes, highlight and describe type of injury, age when noted, consults, test.

**Neonatal Feeding problem**
___ No ___ abnormal or difficulties: length of time ___ days ___ MI

**Neonatal Respiratory disorders**
Respiratory conditions continuing beyond initial resuscitation or those that developed after the first 20 minutes of life: ___ Yes ___ No ___ MI
If yes, highlight and summarize age when noted, consults, test results.

**Neonatal non-neurological changes** (other abnormal conditions not previously recorded)
___ Hyperbilirubinemia ___ Phototherapy ___ other
If yes, highlight and summarize age when noted, consults, test results.

**Neonatal neurological condition:** abnormal neurological conditions present at birth or those that anytime prior to discharge: ___ Yes ___ No ___ MI
If yes, highlight and record onset time, duration and abnormal summary if neurologist consulted, type of injury, age when noted, consults, tests.

- Tension fontanel: normal abnormal
- Posture reflex: normal abnormal
- Segmental reflex: normal abnormal: clonus DTR’s
- Primitive reflex: normal abnormal: grasp suck Moro
  - ATNR other
- Tone: normal abnormal: hypotonia/floppy hypertonic other
- Level of consciousness:
- Respiratory drive: normal decreased: regular irregular
  - none
- Cranial nerve function: normal abnormal
- Neonatal seizure/ Jitteriness: yes no MI 
  - treatment:
- Tests: (summarize of abnormal eg: ultrasound, EEG, CT, MRI)

**Summary:**

**IV. Cerebral Palsy (CP)**

- age: ____ Y ____ M (at time of most recent functional estimation)
- age of diagnosis of CP: ____ Y ____ M
- social service: ____ Yes ____ No If yes, name of service institution:

**4.1. Classification of Cerebral Palsy**

- spastic type: monoplegia hemiplegia paraplegia diplegia __
  - quadriplegia other
- dyskinetic type: athetosis chorea dystonia tremor
  - rigidity other
- ataxic type: congenital cerebellar ataxia other
- atonic type: diplegia congenital (simple) other
- mixed type: predominant type: distribution:
  - unclassified other MI

**4.2. Etiology of CP**

- prenatal factors
  - genetic factors: chromosomal finding
  - Family history:
  - acquired during gestation:
___ confirmed maternal infection ___ irradiation
___ prenatal anoxia: ___ placental abnormalities or
___ maternal anoxia ___ maternal toxemia
___ hemorrhage during pregnancy
___ fetal cerebral hemorrhage
___ predisposition to miscarriage
___ metabolic disturbances during pregnancy
___ small for gestational age
___ other

___ perinatal/neonatal factors
___ hypoxic ischemic encephalopathy:
   APGAR score: 5 min ___ 10 min ___ 20 min ___
   ___ mechanical respiratory obstruction e.g. ___ coiling of cord
   ___ meconium aspiration ___ CPD
   ___ prolonged fetal bradycardia ___ other
   ___ intracerebral hemorrhage ___ malpresentation of labor: ______
   ___ mechanical factors depending upon the type of delivery
   ___ prolong labor ___ CPD ___ other
   ___ periventricular hemorrhage: Grade ___
   ___ confirmed brain edema or evidence of neonatal shock
   ___ sepsis or CNS infection
   ___ other

___ postneonatal factors
___ traumatic injuries ___ infections ___ toxic factors ___ seizure
___ vascular accident ___ cerebral anoxia ___ brain tumors
___ kernicterus ___ other

___ Prematurity or low birth weight
___ low birth weight (1500-2500) ___ very low birth weight (1499-1000g)
___ extreme low birth weight (<1000g), with:
   ___ frequent prolonged apnea
   ___ periventricular hemorrhage: grade ___
   ___ intracerebral hemorrhage ___ chronic lung disease ___ other
___ Unclassified or ___ Idiopathic ___ other ___ MI

4.3. **Severity of CP** (according to Gross Motor Function Classification System)
___ level 1 ___ level 2 ___ level 3 ___ level 4 ___ level 5 ___ MI

4.4. **Manual dexterity**
   ___ (1) normal — upper limbs unaffected.
   ___ (2) mild disability — some clumsiness of fine movements but able to
      feed and dress without assistance.
   ___ (3) moderate disability — able to feed and dress with difficulty.

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(4) severe disability — unable to feed and dress without assistance
(5) MI

4.5. Other functional domain

- Eating skills: __ finger feeding ___ unable to feed by self ___ tubal feeding ___ other __ MI
- Toilet: __ trained ___ partial trained ___ not trained ___ other ___ MI
- Receptive nonverbal: __ communication ___ some ___ none ___ other ___ MI
- Speech skills: __ well expression ___ poor expression ___ can not speech ___ other __ MI
- Recognize language ability: ___ well ___ moderate ___ poor ___ other ___ MI
- Social response: ___ well ___ moderate ___ poor ___ other ___ MI

4.6. Mental ability

- IQ: ___ age of test (Y/M) ___/___ Method: ______
  (1) normal (IQ>85)
  (2) mild learning disability (IQ 70-84)
  (3) moderate learning disability (IQ 50-69)
  (4) severe learning disability (IQ <50)
  (5) too young or suspected severe to profound but is not testable
  (6) MI

4.7. Epilepsy in CP

- Non-existent ___ existent: onset time (Y/M) ___/___ ___ MI
  ___ frequency of seizure: ___ < 1 per day ___ 1 per day ___ other: ___
  ___ type of seizure: ___ generalized ___ partial ___ infantile spasm ___
    partial with secondary generalization ___ unclassified ___ other: _______ ___ MI
- Treatment of seizure: ________
  Outcome of epilepsy after treatment:
  ___ seizure free off medication ___ seizure free on medication
  ___ occasionally seizure on medication ___ frequently seizure on medication ___ other ___ MI

4.8. Feeding problems in CP

- Onset time of feeding problem (Y/ M) ___/___
- Type of feeding problems: ___ difficult feeding ___ difficult swallowing ___ G-E reflux ___ other
- Treatment of feeding problems: ___
4.9. Respiratory problems in CP

___ frequent respiratory infection
___ documented aspiration pneumonia
___ need respiratory equipment supportive treatment
___ documented reactive respiratory disease (e.g.: asthma)
___ other ___ MI

4.10. Orthopaedic problems in CP

___ scoliosis ___ kyphosis ___ dystonia ___ other
managed with:
___ medicine: ___ Baclofen for spasticity ___ other
___ supportive treatment: ___ standing frame ___ posterior walker
___ wheelchair: ___ self propel ___ assistance
___ electric wheelchair
___ night splints to ankles ___ brace
___ soft splints ___ ankle-foot orthoses (AFO)
___ footwear attached with/without below-knee calipers ___ seating systems
___ other
___ surgical treatment: ___ selective dorsal rhizotomy
___ tendon release: ___ hamstring ___ adductor
___ iliopsoas ___ other
___ age of operation: ___ Y ___ M
___ tendon transfer
___ rotation osteotomies ___ other
___ other treatment:

4.11. Visual Impairment

___ cortical visual impairment ___ complete blindness
___ field defect ___ strabismus ___ amblyopia
___ nystagmus ___ other
___ visual dysfunction:
___ mild (vision is > 20/60 in the better eye).
___ moderate (vision is 20/60 to 20/200 in the better eye).
___ severe (vision is < 20/200 in the better eye).
4.12. Hearing Impairment
- complete deafness
- partial deafness
- neurosensory deafness
- other

- hearing loss (hearing loss is average across frequencies 0.5-4 kHz):
  - mild (hearing loss is 21-45 dB).
  - moderate (hearing loss is 45-70 dB).
  - severe (hearing loss is > 70 dB).

4.13. Other
- other infectious problems:
- any other major medical condition:

4.14. Diagnosis (Total disorders, associated with CP) Y M

4.15. Management of CP
- the time of diagnosis as CP (D/M/Y): ___/___/___
- age of start of treatment (Y/M): ___/___
- location of care: ___ at home ___ institution ___ hospital
  ___ foster family ___ alternative home care
  ___ other ___ MI
- type of treatment: ___ early intervention
  ___ medicine
  ___ rehabilitation
  ___ surgical procedure
  ___ type of operation:
  ___ other ___ MI

4.16. Outcome
- alive ___ deceased ___ resolutive ___ loss of follow-up
- prominent problems:
  ___ respiratory problem
  ___ feeding problem
  ___ orthopedic problem
- date of death (D/M/Y): ___/___/___
- time (Hr : Min): ___ : ___
- age of death (M/Y): ___/___
- cause of death:
4.17. Summary: