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Scoliosis in cerebral palsy

KATINA PETERSSON

FACULTY OF MEDICINE | LUND UNIVERSITY



Scoliosis in cerebral palsy

Children with cerebral palsy have an increased risk of developing scoliosis, which can affect their quality of life negatively. The purposes of this thesis were to describe the development of moderate and severe scoliosis in individuals with CP, to identify predictors for severe scoliosis and to create a risk score based on those predictors. We have also explored the use of spinal orthoses in a total population of individuals with CP and evaluated an instrument to examine health related quality of life in children with low gross motor function.



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She started her PhD-studies at the Department of Orthopaedics, Lund University, in 2013. She was granted funds in 2015 from the Norrbacka-Eugenia foundation, and was a full-time doctoral student between 2016 to 2019.

Katina Pettersson was affiliated to, and worked at the Centre for Clinical Research Västerås, Västmanland County Hospital, during her doctoral studies.



Scoliosis in cerebral palsy

Katina Pettersson



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DOCTORAL DISSERTATION

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Title and subtitle Scoliosis in cerebral palsy		
Abstract		
<p><i>Background</i></p> <p>Individuals with cerebral palsy (CP) have an increased risk for scoliosis, especially those with low gross motor function, affecting both quality of life and overall function. The aims were to increase the knowledge on the development of, and predictors for scoliosis and the use of spinal orthoses in individuals with CP, and also to evaluate the Caregiver Priorities and Child Health Index of Life with Disabilities (CPCHILD) questionnaire to examine health-related quality of life (HRQoL).</p> <p><i>Method</i></p> <p>The CPCHILD was distributed to 123 families and analysed for validity and test–retest reliability (<i>Study I</i>). Studies II–IV were based on registry data from the Cerebral Palsy Follow-up Programme (CPUP). Incidence was analysed for scoliosis related to age, sex and level of gross motor function according to the Gross Motor Function Classification System (GMFCS) for 962 individuals born 1990–2012 in southern Sweden (<i>Study II</i>). The use of spinal orthoses was analysed for 2800 children aged 1–14 years in relation to age, sex, GMFCS level, degree of scoliosis, treatment goals and goal attainment (<i>Study III</i>). The risk for developing severe scoliosis after 5 years of age and before the age of 16—with predictors based on risk factors at the age of 5—was analysed for 654 children with CP (<i>Study IV</i>).</p> <p><i>Results</i></p> <p>The CPCHILD showed good construct validity and ability to discriminate between GMFCS levels, and test–retest reliability was high for total and domain scores (<i>Study I</i>). The number of people with scoliosis increased up to 20–25 years of age, and incidence was related to age, sex and GMFCS level (<i>Study II</i>). The use of spinal orthoses increased with age and GMFCS level, functional goals were most common, and goal attainment was high (57–87%) (<i>Study III</i>). The predictive ability of the risk score was high with an area under the curve value of 0.874 (<i>Study IV</i>).</p> <p><i>Conclusion</i></p> <p>Surveillance programmes for scoliosis in CP should be based on age, GMFCS level and should be initiated at a young age and continued into adulthood. The individual risk factors can help to initiate and implement preventive interventions and strategies at an early stage. Children with postural deficits with or at risk for scoliosis should be given the opportunity to explore the functional benefits of a spinal orthosis. The CPCHILD appears to be a valid and reliable proxy-reported measure for HRQoL in children with CP.</p>		
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Scoliosis in cerebral palsy

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MADE IN SWEDEN 

Per aspera ad astra
Through difficulties to the stars

Jag ska bara...Alfons Åberg

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Original papers

This thesis is based on the following original papers and are referred to in the text by their Roman numerals:

- I Pettersson K, Bjerke KM, Jahnsen R, Ohrvik J, Rodby-Bousquet E. Psychometric evaluation of the Scandinavian version of the caregiver priorities and child health index of life with disabilities. *Disabil Rehabil.* 2019;41(2):212–8.
- II Hägglund G, Pettersson K, Czuba T, Persson-Bunke M, Rodby-Bousquet E. Incidence of scoliosis in cerebral palsy. *Acta Orthop.* 2018;89(4):443–7.
- III Pettersson K, Rodby-Bousquet E. Prevalence and goal attainment with spinal orthoses for children with cerebral palsy. *J Pediatr Rehabil Med.* 2019;12(2):197–203.
- IV Pettersson K, Wagner P, Rodby-Bousquet E. Development of a risk score for scoliosis in children with cerebral palsy. (Submitted)

Abbreviations

AUC	Area under the Curve
CI	Confidence Interval
CP	Cerebral Palsy
CPCHILD	Caregiver Priorities and Child Health Index of Life with Disabilities
CPUP	Cerebral Palsy Follow-up Programme and National Quality Registry
GMFCS	Gross Motor Function Classification System
HR	Hazard Ratio
ICC	Intraclass Correlation Coefficient
MP	Migration Percentage
OR	Odds Ratio
ROC	Receiver Operating Curve
ROM	Range of Motion
SCPE	Surveillance of Cerebral Palsy in Europe

Definitions

Cerebral palsy	A group of permanent disorders of the development of movement and posture, causing activity limitations, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of CP are often accompanied by disturbances of sensation, perception, cognition, communication, and behaviour, by epilepsy and by secondary musculoskeletal problems ^{1,2} .
Hip dislocation	Lateral displacement of the femoral head, with a migration percentage of 100% ³ .
Posture	The shape of the body. The anatomical alignment of the body segments in relation to each other and the supporting surface; also, the relationship between the body and the environment.
Postural ability	The ability to stabilize the body segments relative to each other and to the supporting surface and to achieve the most appropriate body configuration for the performance of the particular task and environment. This means control of the centre of gravity relative to the base of support during both static and dynamic conditions.
Mobility	Transferring from one place to another, in any kind of form of transportation.
Scoliosis	A lateral deviation of the spine in the frontal plane.
Risk score	A score for the individual risk to develop scoliosis before the age of 16 years, based on four independent risk factors at the age of 5 years: GMFCS levels IV and V, female sex, epilepsy and having limited knee extension.

The thesis at a glance

Study	Questions	Methods	Results	Conclusions
I	What are the psychometric properties of the Scandinavian version of the CPCHILD for children with CP?	123 families, with children with CP, in Sweden and Norway agreed to evaluate the CPCHILD for test–retest reliability and construct validity.	The CPCHILD showed high known-group validity ($p < 0.001$), high test–retest reliability (ICC 0.92) and high internal consistency ($\alpha = 0.96$).	The CPCHILD is a reliable and valid health-related proxy-reported questionnaire for children with CP.
II	What is the incidence of scoliosis in a total population of individuals with CP? What is the association between scoliosis and GMFCS, age and sex?	Prospective cohort study of all 1025 individuals with CP born 1990–2012 in southern Sweden, based on clinical and radiographic examination of the spine.	The incidence of scoliosis increased with GMFCS level and age up to 20–25 years.	Surveillance programmes for scoliosis should be based on age, GMFCS level from an early age and continue into adulthood.
III	What are the treatment goals and goal attainment for spinal orthoses for children with CP? What are the associations between treatment goals and goal attainment for spinal orthoses and age, sex, GMFCS level and scoliosis?	Cross-sectional study of a total population ($N = 2800$) of children with CP. Treatment goals and goal attainment were based on the four CPUP goals: 1) prevent deformity; 2) improve stability; 3) improve arm–hand function; and 4) improve head control.	Spinal orthoses were used by 9%, and increased with age and GMFCS level. Spinal orthoses were mainly used to improve function and goal attainment was 78–87% for functional goals.	Children with postural deficits should be given the opportunity to explore the functional benefits of a spinal orthosis.
IV	What are the predictors for scoliosis in 5-year-old children with CP? Can the predictors be used to develop a risk score to predict severe scoliosis before the age of 16?	Prospective register study of 654 children with CP born 2000–2003, based on the examination closest to 5 years of age. Eight potential predictors were analysed for scoliosis at the age of 16.	Female sex, GMFCS IV and V, epilepsy and limited knee extension were significant risk factors. The predicted ability of the risk score was high (AUC=0.86).	The risk score for scoliosis may be useful when considering interventions to prevent or predict severe scoliosis in young children with CP.

It all starts with a patient

As for all of us who work with patients, it only takes one patient to start your journey towards knowledge—and this is my patient; let’s call him “Kid”.



Kid is a 10-year-old boy who came to Sweden as a refugee in 2012. At the habilitation centre, he was assigned to me. He was classified with a bilateral spastic cerebral palsy (CP), and low level of gross motor function (GMFCS level V), without mobility options and difficulties with stability, head control and arm-hand function. In his homeland, there was little help for disabled children. Finally, in Sweden, Kid got a new wheel-chair, but it was not adapted for his needs and he sat poorly. Because of low muscle tone and muscle endurance, he needed more stability than we could offer with this wheel-chair. It was decided that he needed a spinal orthosis, together with a customized wheel-chair. At the time, he had no scoliosis, but with a slumped seating posture sustained over long periods of time, there was a high risk that he would eventually develop a spinal deformity.

All questions addressed in this thesis can apply to Kid’s problems: he had a poor sitting position, with risk for scoliosis. *How many children and adults with CP have scoliosis?* (Study II). Kid needed a brace for better posture in sitting. *How many children wear a brace? What are the treatment goals and goal attainment?* (Study III). Kid was a patient at risk for scoliosis. *Can we predict his individual risk for scoliosis as a teenager?* (Study IV). What did Kid and his family think about the interventions given by us? *How could we examine Kid’s health-related quality of life, which was at GMFCS level V?* (Study I).

Att inse att man är okunnig, är ett bra steg mot kunskap
Benjamin Disraeli 1804–1881

Introduction

Cerebral palsy (CP)

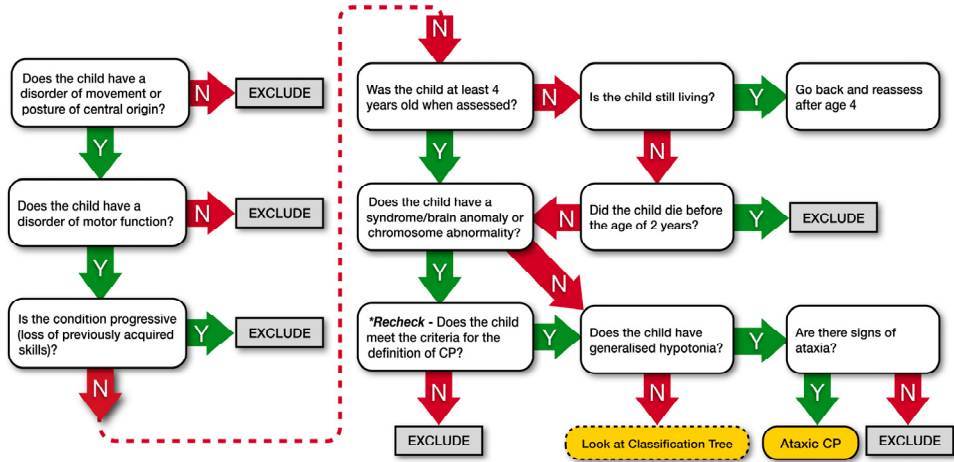
In 1843, William John Little, an English orthopaedic surgeon, published a series of lectures in which he described different musculoskeletal deformities⁴. In 1862, he presented his seminal work on deformities. He differentiated between congenital deformities observed at time of birth and limb deformities that developed after early, difficult or traumatic births into what he called spastic rigidity⁵. He presented 47 cases and grouped them into three categories: 1) hemiplegic rigidity; 2) paraplegia affecting both legs more than the arms; and 3) generalized rigidity. What he described was then called “Little’s disease”, today more commonly known as CP.

Definition of CP

CP is a heterogeneous disorder that reflects the clinical descriptive signs and can be considered more of an umbrella term^{6,1}. The definition mostly used today is that of Rosenbaum and colleagues¹ (p. 9); “Cerebral palsy (CP) describes a group of permanent disorders of the development of movement and posture, causing activity limitations, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication, and behaviour, by epilepsy, and by secondary musculoskeletal problems”.

With respect to the definition of CP, the Surveillance of Cerebral Palsy in Europe (SCPE)—a collaboration of CP surveys and registers—has inclusion and exclusion criteria that rest upon five key points: 1) an umbrella term; 2) permanent but not unchanging; 3) involving a disorder of movement and/or posture and of motor function; 4) due to a non-progressive interference, lesion or abnormality; and 5) the interference, lesion, or abnormality is in the immature brain, generally considered to be before the age of 2 years⁷. SCPE provides clinicians with a decision flow chart for inclusion and exclusion criteria of CP, to aid classification into neurological and topographical categories (Figure 1)^{7,8}.

Decision tree for Cerebral Palsy



SCPE Collaborative Group. Surveillance of cerebral palsy in Europe: a collaboration of cerebral palsy surveys and registers. *Developmental Medicine and Child Neurology*. 2000;42:816-24.

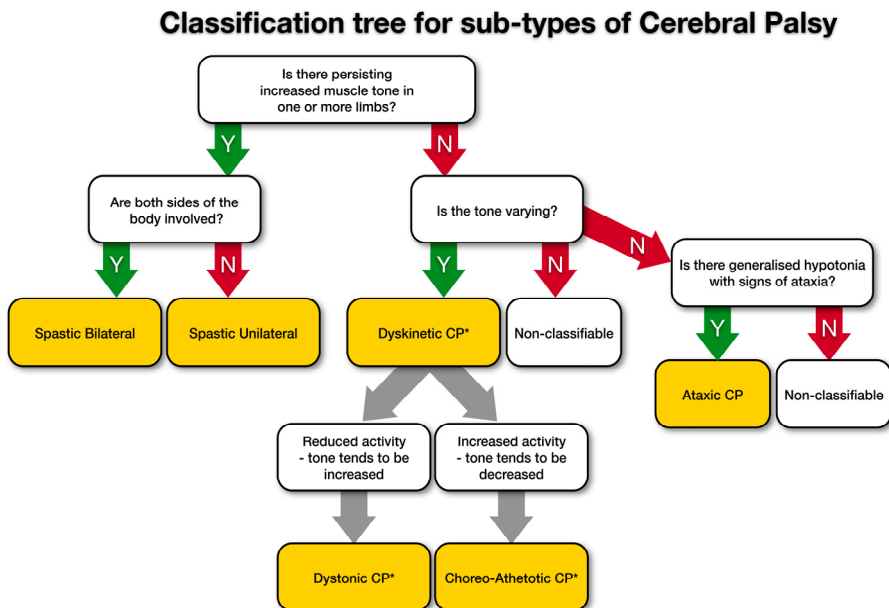
Figure 1. The SCPE decision tree for inclusion or exclusion of cases of CP. The figure is reprinted by permission of SCPE (7).

CP is the most common cause of motor disability in children and adolescents with a prevalence of 2.4–2.7/1000 live births^{9,10}. In 2018, the prevalence in Swedish children aged 5–16 years (born 2001–2012) for CP was 2.16/1000¹¹, and for young adults aged 17–20 years, it was 2.3/1000 in 2013¹².

CP can be one of the most severe disabilities in childhood⁷, beginning in early childhood and persisting through the lifespan¹. CP makes heavy demands on health, educational and social services as well as on families, children and adults themselves^{1,6}. A large registry study from Norway¹³ showed that a majority of the individuals with CP had one or more comorbidities and that the risks of medical, neurological and mental and/or behavioural disorders were considerably higher compared with the general population. However, with medical advances, the probability of survival has increased, even among children with a severe level of disability^{7,14}.

Classification of CP subtypes

Since the nineteenth century, the medical society has debated a universally accepted classification of CP¹⁵, and its classification has changed over time with increasing medical knowledge^{16,17,15}. For many years, the classification by Hagberg et al. was used in Sweden¹⁸. But since the beginning of the twenty-first century, the classification by SCPE has been used by registers and databases that record and describe children and adults with CP⁷. Hagberg et al. categorized CP into spastic uni-, bi-, tetraplegia, ataxic, dystonic or choreo-athetotic CP, or mixed form¹⁸, while the SCPE groups spastic bi- and tetraplegia into spastic bilateral CP. In addition to the decision flow chart for inclusion and exclusion criteria of CP, the SCPE also provides with a hierarchical classification tree (Figure 2)^{7,8}. The SCPE classification has been used throughout Studies I–IV.



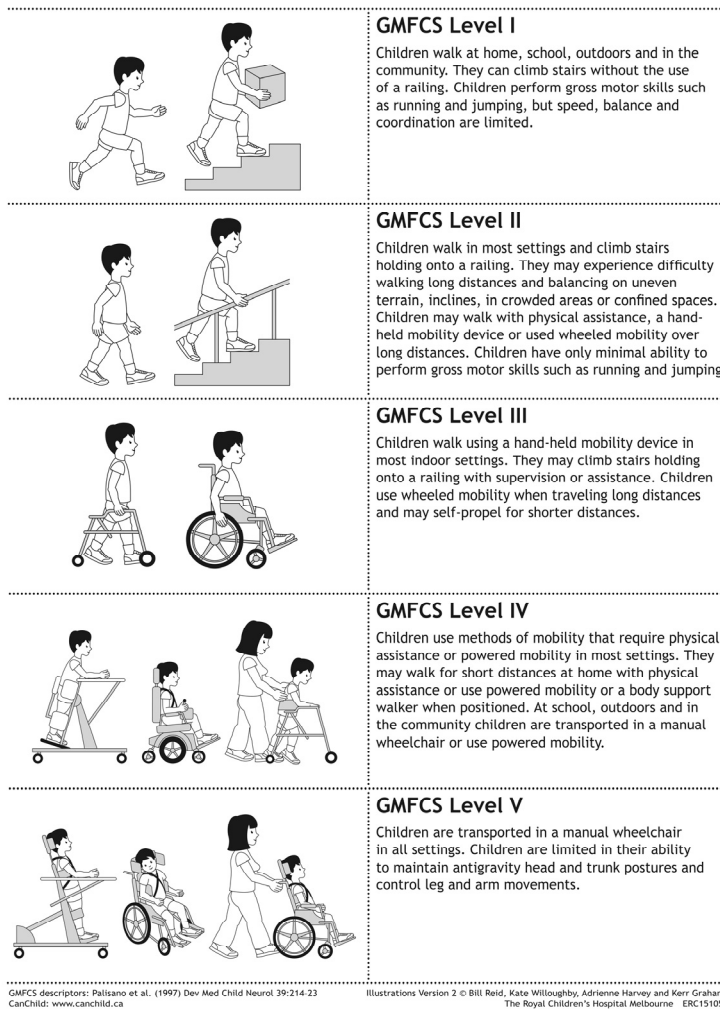
SCPE Collaborative Group. Surveillance of cerebral palsy in Europe: a collaboration of cerebral palsy surveys and registers. *Developmental Medicine and Child Neurology*. 2000;42:816-24.

Figure 2. The SCPE hierarchical classification tree of CP subtypes. The figure is reprinted by permission of SCPE (7).

Classification of gross motor function

In addition to the classification of CP subtypes, Palisano and colleagues¹⁹ developed a standardized system that describes the functional motor ability in children with CP (in age bands from 2 to 12 years) in five levels, known as the Gross Motor Function Classification System (GMFCS) (Figure 3).

GMFCS E & R between 6th and 12th birthday: Descriptors and illustrations



GMFCS descriptors: Palisano et al. (1997) Dev Med Child Neurol 39:214-23
CanChild: www.canchild.ca

Illustrations Version 2 © Bill Reid, Kate Willoughby, Adrienne Hanney and Kerr Graham,
The Royal Children's Hospital Melbourne ERIC151050

Figure 3. The Gross Motor Function Classification System (GMFCS), levels I to V, for ages 6–12 years. The figure is reprinted by permission of Professor H. Kerr Graham.

It was later revised and expanded with an age band from 12 to 18 years²⁰. The GMFCS is classified on a five-point ordinal scale, where level I describes the highest level of function and level V the lowest (Figure 3)²⁰. The emphasis in the GMFCS is on self-initiated mobility such as sitting and walking, and on the use of assistive devices such as walkers and wheel-chairs²¹, with activities that are meaningful for individuals with CP in their daily life²². The child's GMFCS level generally does not change but tends to remain stable; levels I and V are least likely to be reclassified during childhood²²⁻²⁴. The benefit of the GMFCS is its ability to objectively classify children and adolescents with CP and thereby provide support in goal setting, to give support when designing services and interventions and in translating research into practical use²³.

CPUP

The Cerebral Palsy Follow-up Programme (CPUP) started in the south of Sweden in 1994²⁵. Orthopaedic departments and habilitation centres together defined common goals towards detecting and preventing hip dislocations, contractures and deformities in children with CP, with the overall goal being to reduce pain and improve quality of life (QoL). CPUP also aimed to increase knowledge about CP, and to improve co-operation between different professionals working with individuals with CP and their families (www.cpup.se). The Swedish CPUP was designated as a National Quality Registry in 2005 by the Swedish National Board of Health and Welfare.

CPUP has a coverage rate of over 95%, representing almost all children with CP in Sweden⁹. Since 2009, CPUP has also included adults with CP, and at present, over 1600 adults participate¹¹. The incidence of CP among adults of different ages in CPUP is unknown. In 2018, CPUP followed 0.74/1000 inhabitants in the age group 19–27 years (born 1990–1998)¹¹ (p. 11).

Children are enrolled in CPUP at the earliest suspicion of CP, usually at 1–2 years of age. This means that children younger than 4 with suspected but not yet confirmed CP are included in the registry. The diagnosis is verified by a neuropaediatrician from the age of 4, and those not diagnosed with CP are excluded from the registry⁹. Exclusion and inclusion criteria are in accordance with those of the SCPE (Figure 1)⁷.

As a result of CPUP, a 10-year follow-up showed that the incidence of hip dislocations was reduced from 8.0% to 0.5% in Sweden, and with continued low figures during the 20-year follow-up^{26,27}. Through early detection and timely management, severe contractures and scoliosis were also significantly reduced^{28,27,29-31}, together with the need for surgery for severe contractures²⁸.

Other countries have adopted CPUP and this standardized surveillance programme is used in Norway (since 2002), Denmark (2010), Scotland (2012) and parts of Iceland and Australia (both 2012). Planning is in progress to implement CPUP in several other countries.

Clinical examination in CPUP

CPUP includes a continuous standardized follow-up of radiographic examinations of hips and spine, clinical examination of dominant neurological symptoms, gross and fine motor function, measurements of range of motion (ROM), spasticity assessments, spinal assessments, posture, mobility and reports of pain and physical activity. Treatment that the child or adult receives is also reported, for example, the use of orthoses, including spinal orthoses, spasticity-reducing treatments or surgical treatments. All measurements are reported by each therapist to the web-based CPUP database ²⁵. The individuals with CP are examined by their local physiotherapist (PT) and occupational therapist (OT) following standardized protocols (<http://cpup.se/in-english/manuals-and-evaluation-forms/>). The examinations are done at different intervals related to age and GMFCS level (Figure 4) ²⁵.

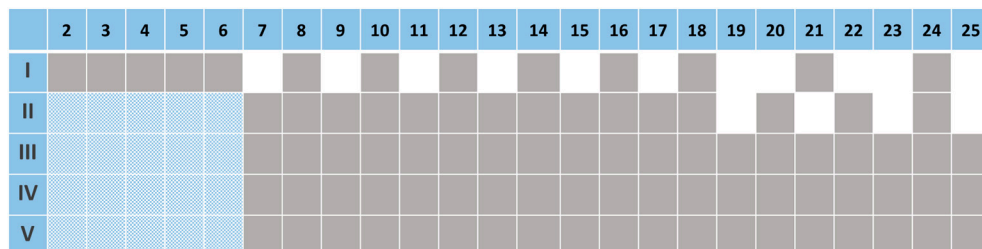


Figure 4. Standardized follow-up related to age and GMFCS level following examination by a physiotherapist and an occupational therapist. Grey: examination once a year; Blue striped: examination twice a year.

The PT examination includes a spinal assessment that is performed with the child in a sitting position on a plinth, both in a forward bend and in an upright position. The spine is graded as follows.

No scoliosis	no visible curve
Mild scoliosis	discreet curve visible only on a thorough examination during forward bend
Moderate scoliosis	obvious curve visible during both in the upright position and forward bend
Severe scoliosis	pronounced curve preventing the child from attaining an upright position without external support

The psychometric properties of the clinical examination show high concurrent validity compared with radiographic Cobb angle, with a sensitivity of 75%, specificity of 95.8% and excellent interrater reliability ($\kappa=0.96$)³².

The clinical spinal assessment is used as a screening method to identify children in need of radiographic follow-up. According to the programme, children with mild scoliosis are only monitored clinically, regardless of age and GMFCS level. Children under 8 years of age with flexible, moderate scoliosis are also only followed clinically, while those with a rigid, moderate scoliosis or a severe scoliosis, are examined radiographically in a sitting position (frontal and sagittal). Children older than 8 with a moderate or severe scoliosis are examined radiographically (www.cpup.se).

Indications for considering surgical treatment are as follows.

- a) Scoliosis with a Cobb angle $>40^\circ$
- b) Scoliosis $>30^\circ$ with a rigid pelvic obliquity ($>10^\circ$)
- c) Scoliosis $>30^\circ$ with pelvic obliquity and a migration percentage $>40\%$

The spine in CP

In ancient Greece, the “father of medicine” Hippocrates (460–370 BC) (Figure 5) described *spina luxate*, in which he included all vertebral deviations. Hippocrates was the first in medical history to write a systematic presentation of both the anatomy and pathology of the spine. He wrote 60 books and information about spinal deformities was included in several of them, but especially in “The Book on Bones”³³. Scoliosis derives from the Greek word *scolios* meaning crooked³⁴.

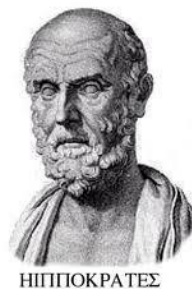


Figure 5. The “father of medicine” Hippocrates.

Scoliosis is a diverse group of conditions that consists of changes in the shape and position of the spine, thorax and trunk ³⁵. When the normal spine is looked at in a frontal plane, it seems straight, even though the spine is curved in the sagittal plane, with a cervical and lumbar lordosis and thoracic kyphosis ³⁴. It occurs in 2–3% of apparently healthy children, called idiopathic scoliosis ³⁵. The term neuromuscular scoliosis describes a non-congenital spinal deformity that occurs in patients with any type of pre-existing myopathic or neuropathic diagnosis, such as in children and young individuals with CP ^{36,37}. The focus in this thesis is on neuromuscular scoliosis, hereafter referred to as simply scoliosis. Scoliosis is traditionally defined as:

“A lateral deviation of the spine in the frontal plane

Radiographic evaluation is created by an antero-posterior view of the entire spine, preferably in a weight-bearing position. The lateral deviation is often accompanied by a rotation and wedge formation of the vertebrae, which makes scoliosis a three-dimensional spinal deformity ³⁴. The Cobb angle is the most commonly used method to measure the degree of scoliosis, determined by the most tilted vertebrae at each end of the curve (Figure 6) ³⁸.

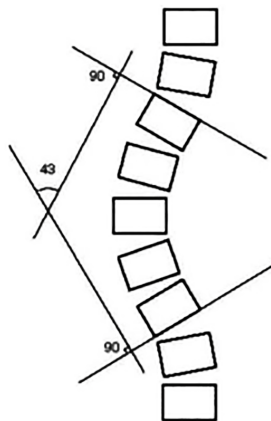


Figure 6. Measurement of the Cobb angle.

To calculate the Cobb angle, the vertebrae with the maximally tilted end-plates below and above the apex on the radiographs are identified. Erect intersecting perpendiculars from the superior surface of the top and the inferior surface of the bottom vertebrae of the curve and the two intersecting perpendiculars form the scoliosis angle³⁹. It is important to recognize following significant points regarding measurement of the Cobb angle: it is a two-dimensional measurement of a three-dimensional condition and the angle can change if different vertebrae are used to measure it.

A scoliosis may consist of an elongated C-shaped curve, as manifested in the majority of children with CP^{40,41}, or an S-shaped curve, including a primary and secondary curve^{42,43,41}, with or without pelvic obliquity (Figure 7)^{44,40}.

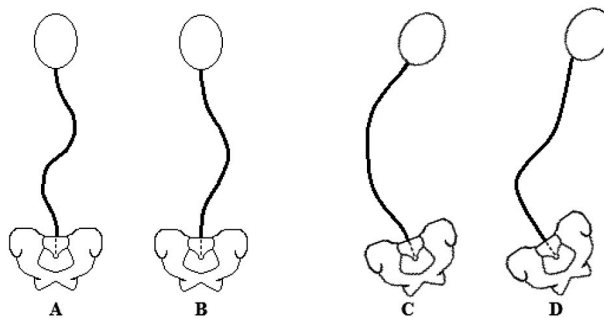


Figure 7. Different spinal deformity patterns in neuromuscular scoliosis. In types A and B, the spine is well balanced due to the double curves, with little or no pelvic obliquity. Types C and D comprise large thoracolumbar or lumbar curves that are more C-shaped, and the deformity extends into the sacrum, thereby causing pelvic obliquity.

Aetiology of scoliosis

The Hueter–Volkman law

Two articles were published in Germany in 1862 independently by the surgeons Carl Hueter and Richard von Volkmann. They stated that the growth of bones is accelerated by reduced compression (Hueter), and inhibited by compression (Volkmann). In a later article, Volkmann linked these two statements together, thereby forming the Hueter–Volkman law that still persists today⁴⁵. The law helps us understand the mechanical modulation of epiphyseal growth in all immature bones^{45,46}. The reasons why this regulation of growth occurs are complex, but genetic, vascular, hormonal and biomechanical factors all contribute⁴⁷. When applying asymmetric compression forces on a vertebrae, wedging can develop due to a combination of asymmetrical growth, vertebral body remodelling and epiphyseal wedging⁴⁸, thereby giving rise to the “vicious cycle” of scoliosis progression (Figure 8)⁴⁶.

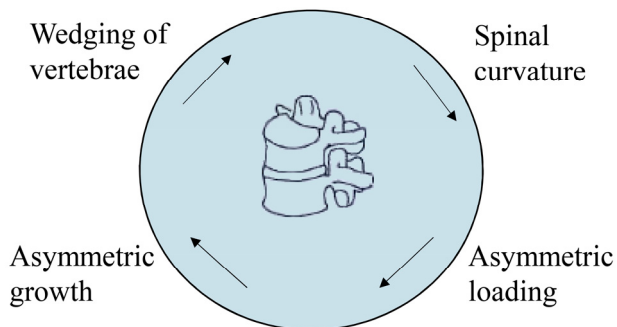


Figure 8. The "vicious cycle" of scoliosis progression, illustrating how scoliosis, depending on where the problem starts, continues to develop.

The wedging of discs and vertebrae develops with increasing progression of scoliosis, with maximum wedging at the apex of vertebrae⁴⁹. Wedging of either disc or body depends on the location, with higher disc wedging in thoracic areas and higher vertebral wedging in lumbar or thoracolumbar areas^{49,50}. For the development of wedging, animal studies have shown that asymmetric growth is more important in younger animals, and vertebral remodelling is more important in older animals. There may be implications of this knowledge for treatment also in humans⁴⁸.

Wolff's law

In 1892, German orthopaedic surgeon Julius Wolff published a book entitled "The Law of Transformation of Bones"⁵¹⁻⁵³. Wolff's law describes that bone is in constant remodelling in response to prevailing mechanical demands through time, with internal remodelling as well as possible alteration of external shape. This law of vertebral body remodelling also contributes to the understanding of vertebral wedging in scoliosis⁴⁸, together with the principles of the Hueter–Volkman law.

The cause of scoliosis and which component of the neurological deficit is more responsible are not entirely clear. A combination of incomplete muscle control and muscle weakness, hyper- and hypotonus, poor balance and especially truncal imbalance are all contributory factors⁵⁴⁻⁵⁶.

Incidence and risk factors

Scoliosis is common in non-ambulatory individuals with CP^{57,58} and children and adults with CP are at high risk for developing scoliosis compared with individuals without CP. Scoliosis can give rise to significant problems depending on age, neurological subtype and GMFCS level^{58,30,31}.

Different definitions of scoliosis, age groups and distribution of gross motor function have led to variations in the reported incidence of scoliosis in people with CP^{59,58,55}. In Sweden, the prevalence ranges from 11% in children to 29% in young adults with CP³⁰, but with a general reported incidence of 20–25%^{59,41,57,60} that can increase up to 64–72% in individuals with total body involvement^{57,41,61}.

The debut for scoliosis often occurs around 10 years of age⁵⁷, but can in many cases start earlier³⁰. Scoliosis with a Cobb angle $>40^\circ$ at the age of 15 progresses in 85% of cases⁵⁷. For children at GMFCS levels I or II, there is a low risk of developing neuromuscular scoliosis³⁰, similar to that of adolescent idiopathic scoliosis in typically developed children⁶².

Information on the risk factors for development of scoliosis in children with CP⁶³ is ambiguous. Age^{30,31} and early onset of scoliosis have been identified as risk factors for severe scoliosis^{57,64,30,65}. Low gross motor performance, at GMFCS levels IV or V, has also been identified as risk factors for scoliosis in children with CP^{63,30}. Recent work suggests that girls with CP have a higher risk than boys for developing scoliosis⁵⁶. The following potential risks factors have also been suggested to contribute to the development of scoliosis. Epilepsy is an independent risk factor associated with scoliosis^{56,66}. Lateral displacement of the hips, hip dislocations, and previous hip surgery have been associated with neuromuscular scoliosis^{30,56,31}, whereas successful hip surveillance leading to a reduced number of dislocated hips results in a lower proportion of scoliosis²⁷. In addition, limited hip or knee extension is associated with scoliosis, windswept hip deformity and postural asymmetries in adults with CP^{12,67}.

Clinical implications and management

Severe scoliosis not only affects the spine but also has a great impact on the individual's quality of life and factors such as posture and stability, including sitting ability, transfer and mobility, as well as care needs and pulmonary function^{34,68}. Thereby, scoliosis also has a great impact on the family and close environment. Pain due to scoliosis can be a problem and pain intensity increases with degree of scoliosis⁶⁹. Orthopaedic problems like pelvic obliquity, dislocated hips and deformities like windswept hips can also develop with greater curves^{70,71}. Respiratory and cardiac complications can occur without interventions, in the worst-case scenario, scoliosis can lead to premature death⁵⁷. The severity of CP in

terms of gross motor function is related to the rate of progression and severity of the curvature^{57,31}.

In ancient Greece, the treatment options for scoliosis included hydrotherapy, physiotherapy, hygienic rule, diet, drug therapies, and minor surgical procedures³³. The first definition of orthopaedics in the eighteenth century highlights the importance of “conservative treatment” rather than using “surgery” concerning children⁷².

Today, the overall goal of management is directed at maintaining or improving functional abilities like seating and positioning, to ease daily care and to reduce pain in order to optimize QoL. The treatment should be tailored to the individual patient, with a treatment plan that includes a detailed risk–benefit assessment based on the severity of coexisting medical morbidities^{54,55,36}. Close surveillance and evaluation are key components for identifying curve progression and improving or maintaining overall function^{28,26,27}. Non-operative treatment options consist of regular clinical assessments of the spine and the use of spinal orthoses^{73,74}, but may also include adaptive seating⁷⁵ and postural management^{55,54}. Surgery may be the definitive treatment option for scoliosis^{76,77}.

Suitable postural support is vital for young children as well as adults when unable to or when experiencing difficulties to maintain or change a position^{67,12,78,79}, and lack of stability is often the main indication for a support like a spinal orthosis^{74,73}. Spinal orthoses may provide functional benefits through postural support that preserves and improves functional abilities^{77,80,74,73,81}. Abilities such as stability⁷⁷, and improved head^{73,81} and hand control^{74,73} are all vital for the QoL of children with CP.

More than 30% of children with CP are non-ambulant, and spend most of their lives in sedentary positions like sitting or lying⁸. In one study, in a population of adults with CP and untreated scoliosis, >50% were non-ambulant⁷⁰. To be non-ambulant increases the risk for scoliosis^{67,12} and contractures, such as knee contractures⁷⁸. A recent study showed that spinal orthoses can give better sitting function, they reduce the need for external support in sitting, and for many children, the number of special adaptations of the chair decreases⁸². The greatest improvements were seen for those who needed support compared with those who could sit freely; however, those children who could sit freely also benefitted from a spinal orthosis⁸². A notable example of the importance of quality in sitting, is that increased comfort was associated with higher QoL⁸³, and that improved sitting balance affected caregiver satisfaction the most^{84,81,77,85}. Although spinal orthoses appear to be effective for the treatment of adolescent idiopathic scoliosis⁸⁶, the results for neuromuscular scoliosis are unclear^{74,77,80}. The main indication for spinal orthoses in individuals with CP, is often to stabilize and delay the progression of spinal curvature and improve stability in sitting^{80,74}. However,

there is a lack of knowledge regarding the treatment goals and the level of goal attainment for the use of spinal orthoses in children with CP ⁸⁷.

Theory of motor control

Motor control can broadly be defined to include both movement, balance and posture. The numerous motor control theories regarding assessment and treatment all combine various elements into one, that is, a systems theory approach. This approach shifts the focus from “normalizing” the individual to normalizing the environment with, for example, assisted devices based on the individual’s needs. The basic core of the systems theory approach is that the interaction of multiple processes, including perceptual, cognitive and motor processes, causes movement to occur, together with the interaction between the individual, the task and the environment (Figure 9). These interacting components make up a systems theory approach that serves as a foundation for many clinical interventions ⁸⁸.

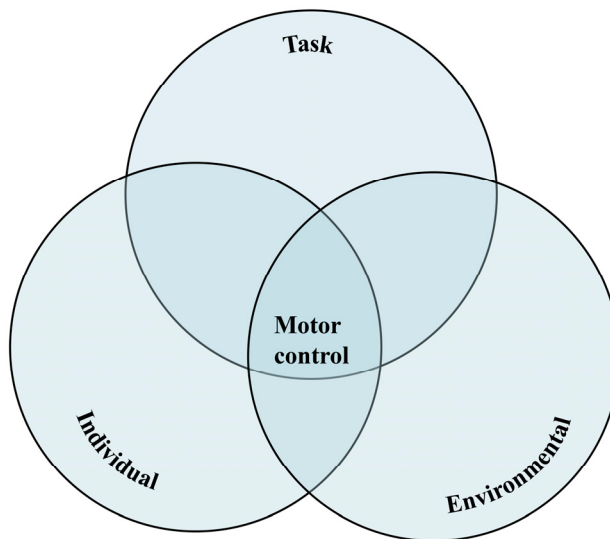


Figure 9. Motor control emerges from an interaction between the individual, the task and the environment.

Postural asymmetries

In 1741, the French paediatrician Nicolas Andry (1658–1742) wrote his pioneering work “L’orthopedie”. Andry also coined the word orthopaedia, made up of the Greek words orthos (straight) and paidos (child). Today’s meaning has expanded to include adults ⁷².

Movement and posture are key problems for children with CP ⁸⁹. However, just as is the case in most children, the newborn child with CP usually has no deformities or musculoskeletal abnormalities. Even though the cerebral lesion is static, the musculoskeletal pathology is often progressive.

Musculoskeletal deformities may appear in young individuals with low functional abilities, secondary to immobility and not due to the primary neurological pathology itself ⁹⁰. Postural deformities start as a response to the prolonged time the child spends in a preferred position ^{91,79}. A preferred lying posture during the first 12 months of life can give rise to an asymmetrical posture that in time becomes established as tissues adapt due to gravity and biomechanical forces ^{79,90}. Those who are non-ambulant are especially at risk of early development and rapid progress of asymmetrical postural problems ^{91,79}. In a study of young adults, all GMFCS levels were associated with contractures, deformities and inability to change position, and for those with the lowest gross motor function (GMFCS levels IV and V), half could not change position independently and had only one sleeping position ¹². Ágústsson and colleagues found that adults who slept in only one position and could not change position independently had higher odds of both scoliosis and windswept hips than those who could move freely ⁶⁷. With rapid growth during childhood and an inability to deal with the effects of gravity combined with immobility, spasticity and muscle weakness, there is an increased risk of problems such as scoliosis, dislocation of the hip, skeletal deformities and fixed contractures ^{92,87}. This highlights the need for clinicians to properly assess posture from an early age and provide postural support through the lifespan when needed ^{79,12}.

Problems with movement and posture can cause impaired stability ⁸⁹. This is highlighted by the fact that almost 30% of children with CP are non-ambulant ⁸, spending their time sitting or lying, thereby giving larger stability limits ⁸⁹. A lack of stability can affect both head and hand control ⁹³, meaning that the performance of everyday activities is noticeably influenced by postural deficits, depending on the child’s functional level ⁸⁹. Without adequate support, there is an increased risk of developing a functional quadriplegia with reduced ability to use arms and hands ⁵⁵, requiring higher levels of care ⁵⁷.

Contractures are common in CP with knee contractures being the most frequent in the lower extremities for adults with CP, limiting their ability to find optimal lying

and standing positions ¹². Almost 25% in a total population of children with CP had knee contractures, and those with a hamstring angle of less than 120° had a 10-fold increased risk of knee contractures. Maintaining muscle length, especially of the hamstrings, is important for reducing the risk of knee contractures ⁹⁴.

Health-related quality of life (HRQoL) in CP

Quality of life (QoL) is used as a general concept for physical and psychosocial functioning ⁹⁵. The WHO defines QoL as ⁹⁶: “an individual’s perception of their position in life in the context of the culture and value systems in which they live, and in relation to their goals, expectations, standards and concerns”. It can also be conceptualized as a person’s feeling of well-being across many domains, such as physical, social, emotional and spiritual aspects of life ⁹⁵. HRQoL incorporates the individual’s subjective perception of physical and mental health over time ^{97,98}, which is also addressed as a key component in Articles 13 and 14 of the UN Convention on the Rights of the Child ⁹⁹. HRQoL has a focus on the effects of illness and specifically on the impact that treatment may have on QoL ¹⁰⁰.

Assessment tools for HRQoL

The development and use of standardized HRQoL questionnaires have increased during the last decade ^{98,100}. As HRQoL is often seen as a subset of the overall concept of QoL, there is commonly no distinction made between measures of QoL and measures of HRQoL ⁹⁵. It has been proposed that HRQoL measures can be useful in identifying and prioritizing health problems, in following changes in a patient’s health state and detecting responses to treatment, in facilitating co-operation between patients and clinicians, as well as in identifying hidden or unexpected health problems ¹⁰⁰.

The view of HRQoL for people with disabilities has changed over time; nowadays, the individual is asked how he/she perceives their own QoL ⁹⁵. That said, in many cases, proxy reports from either parents or caregivers may be the only solution for assessing HRQoL for people with cognitive disability, impaired communication or for very young children ^{98,101}. It has been shown that parents are better able to estimate their child’s functional difficulties if their child has an active disease, compared with parents of children without any disease. The former are more aware of their child’s special needs that must be addressed ⁹⁸. It can be difficult for clinicians to judge their patients’ HRQoL ¹⁰².

The CPCHILD

The Caregiver Priorities and Child Health Index of Life with Disabilities (CPCHILD) is a disease-specific questionnaire. It was developed in Canada specifically for the evaluation of HRQoL in children with CP with more severe motor and cognitive disabilities (GMFCS IV and V). The CPCHILD is recommended when measuring HRQoL over time and for measuring the effectiveness of interventions intended to improve or maintain HRQoL. The questionnaire is also suitable for children with traumatic or acquired brain injury¹⁰³. The CPCHILD has been translated into several languages and evaluated for its psychometric properties and has shown to be a valid and reliable measure^{104-107,103}.

The questionnaire has 37 items divided into six domains: 1) *Activities of daily living/personal care*; 2) *Positioning, transferring and mobility*; 3) *Comfort and emotions*; 4) *Communication and social interaction*; 5) *Health*; and 6) *Overall quality of life*. Answers are based on the caregiver's rating of perceived difficulties and level of assistance required for the child during the past 2 weeks when performing activities (Figure 10).

							LEVEL OF ASSISTANCE					
							T O T A L	M O D E R A T E	M I N I M A L	S U P E R V I S E D / D	I N D E P E N D E N T	
Consider how each of the following activities is usually performed by/for your child.												
Rate how difficult each of these activities were in the past 2 weeks,												
and choose the level of assistance that was required to help your child perform these activities.												
During the past 2 weeks, how difficult was the following:		<i>Not Possible</i> <i>(Almost Impossible)</i>		<i>Very Difficult</i>		<i>Slightly Difficult</i>		<i>Very Easy</i>		<i>No problem at all</i>		
1. putting on / wearing footwear? (socks, shoes, braces, etc.)	0	1	2	3	4	5	6	0	1	2	3	

In the above example, the task of putting on / wearing footwear was rated as *very easy*, and the child required a *minimal / supervised* level of assistance to put on footwear.

Figure 10. An example of a question and the corresponding answer on the CPCHILD.

The activities are rated on a seven-point ordinal scale ranging from 6 (no problem at all) to 0 (not possible). The level of assistance ranges from 3 (independent) to 0 (total assistance). The scores are summed for each item, domain and total score according to the CPCHILD manual and interpretation guide. The standardized score for each domain and the total score vary from 100 (best) to 0 (worst). The CPCHILD has recently been translated into Swedish and Norwegian. For the Swedish version of the CPCHILD, see Appendix 1.

Aims

The aims of this thesis were to enhance knowledge on how scoliosis develops in individuals with CP, to explore treatment goals and goal attainment with spinal orthoses, to differentiate children at high risk for scoliosis from those with low risk, and to evaluate a questionnaire to assess HRQoL in children with CP.

- Study I* To examine the psychometric properties of the caregiver version of the CPCHILD for use in children with CP in Scandinavia.
- Study II* To analyse the incidence and prevalence of scoliosis in children and young adults with CP in relation to gross motor function, sex and age.
- Study III* To analyse the treatment goals and goal attainment for spinal orthoses in a total population of children with CP in relation to age, sex, gross motor function and degree of scoliosis.
- Study IV* To develop a risk score for prediction of the individual risk of developing scoliosis before the age of 16 years in children with CP.

Materials and methods

Study design

Study I was a psychometric evaluation of the Scandinavian version of the CPCHILD questionnaire for children with CP. Study II was a prospective cohort study describing scoliosis in CP. Study III was a cross-sectional study looking at spinal orthoses. Study IV was a prospective registry study that developed a risk score for scoliosis. Studies II–IV included total populations of children with CP based on data collected from the CPUP registry.

Table 1.
Participants in Studies I–IV.

Demographics	Study I (%)	Study II (%)	Study III (%)	Study IV (%)
Participants	106	962	2800	654
Age, range	3-15	4-25	0-14	4-5
Mean age	7.9	13.5	7.4	4.6
Male /Female	43/ 63	557 /405	1614 /1186	372/ 283
GMFCS I	21 (19.8)	393 (40.9)	1210 (43.2)	264 (40.4)
GMFCS II	25 (23.6)	190 (19.8)	470 (16.8)	117 (17.9)
GMFCS III	21 (19.8)	95 (9.9)	254 (9.1)	53 (8.1)
GMFCS IV	17 (16)	135 (14)	423 (15.1)	115 (17.6)
GMFCS V	22 (20.8)	149 (15.5)	443 (15.8)	105 (16.1)

Participants and methods

Study I

In Study I, the caregiver version (proxy report) of the CPCHILD was used. Families of children with CP born 2000–2011 (n=553) aged 4–15 years from central Sweden and all of Norway were invited to participate in the study. According to our power calculation, a minimum of 10 children per GMFCS level was needed, and four batches were sent with invitations from September 2013 to May 2015.

In total, 123 families agreed to participate, and they lived in both rural and urban settings. Written consent was obtained from all caregivers. The CPCHILD

questionnaire was sent twice to the families (Figure 11). For descriptive data, see Table 1.

To evaluate construct validity, known groups were used, defined as GMFCS levels I–V. For analyses, the first CPCHILD questionnaire from each family (n=106) with $\geq 80\%$ responses per domain was used. Those questionnaires with $>20\%$ missing values for any of the six domains were excluded.

For comparison, validity was also evaluated for the group of questionnaires that were complete with no missing items (n=82) and similar results were obtained for the two groups (Figure 11).

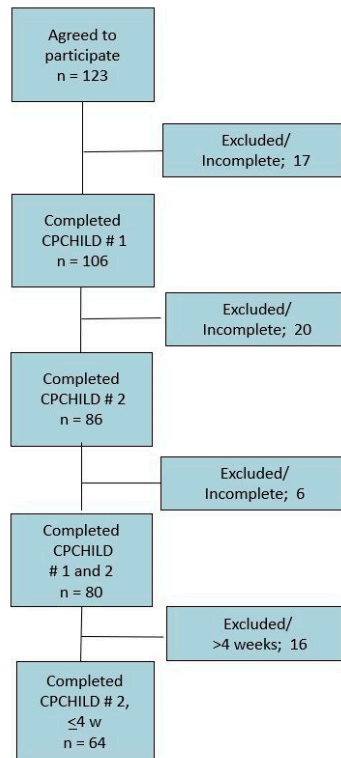


Figure 11. Flowchart for included and excluded/incomplete questionnaires in Study I.

For test–retest reliability, a time span of 2–4 weeks was set for required responses between the first and second questionnaires (n=80). Therefore, analyses were based on the families who answered both surveys with $\geq 80\%$ responses per domain, within the time limit of 4 weeks (n=64) (Figure 11).

Study II

The clinical and radiographic measurements used for the analyses were performed from 1 July 1995 to 3 February 2017 and data were extracted for 1025 children and young adults with CP aged 4–25 years. They were born between 1 January 1990 and 31 December 2012 and were living in southern Sweden (Skåne and Blekinge). Most individuals (n=962) fulfilled the inclusion criteria. Children who died (n=10) or moved out of the area (n=5) before the age of 5 or who moved into the area after the age of 5 (n=58) were excluded. Demographic data are shown in Table 1. The clinical examinations and classification of scoliosis were performed according to CPUP guidelines seen on page 18³². The outcome for Study II was moderate or severe scoliosis. The incidence of scoliosis was analysed in relation to age, sex and GMFCS level.

Study III

Data for 2800 children aged 0–14 years born 2000–2014 were analysed. The latest physiotherapy assessment for each child, performed between 1 January 2013 and 31 December 2014, was extracted and used for analyses. The use of spinal orthoses was analysed in relation to the child's age, sex, GMFCS level and scoliosis. Demographic data are presented in Table 1.

The use of a spinal orthosis was reported as “yes” or “no” for each child. All prefabricated or individually moulded spinal orthoses were included. Treatment goals were set for each child by collaboration between the family and the therapist based on four possible options available: 1) to prevent deformity; 2) to improve stability/positioning; 3) to improve arm–hand function; and 4) to improve head control. Several goals could be reported for each child. In this study, goal 1 focused mainly on body structure, and the prevention of deformity was defined as prevention, reduction or stabilization of scoliosis; goals 2–4 focused more on activities and body function and are hereafter referred to as functional goals. Attainment for each goal chosen was noted as “yes” or “no”; that is, whether or not the child attained the intended goal using the spinal orthosis.

Study IV

The study included national CPUP data of 654 children with CP born 2000–2003. Demographic data are shown in Table 1. We identified predictors in 5-year-old children with CP for development of a severe scoliosis before the age of 16. In this study, scoliosis was defined as either having 1) a radiographically measured Cobb angle of $\geq 40^\circ$, 2) a spinal fusion because of scoliosis, or 3) a severe scoliosis at clinical examination before the age of 16. Mild or moderate curves do not exceed 25° of Cobb angle³² and were treated as no scoliosis. Data from the assessment performed closest to the child's fifth birthday were used for analyses (4.0–5.9 years). The following variables were analysed as potential risk factors for

scoliosis: GMFCS levels IV and V, female sex, spastic subtype, epilepsy, hip surgery, migration percentage (MP) >40%, and limited hip or knee extension by -5° or less.

We grouped GMFCS levels I–III (higher motor function) as the reference category for comparison against GMFCS levels IV and V (lower motor function). Male was used as the reference category for sex^{56,31,108}. Epilepsy was reported as “yes” or “no”, with no epilepsy as the reference category⁵⁶. Neurological subtype was classified as spastic CP (spastic uni- and bilateral CP) versus non-spastic CP (ataxic, dyskinetic and mixed type), and the latter was used as the reference category^{56,66}. All types of hip surgery (including femur osteotomy, pelvic osteotomy and adductor psoas tenotomy) were grouped as hip surgery, with no surgery used as the reference category⁵⁶. Lateral displacement/migration of the hip joint was measured using the MP³, and the highest MP values were used. We defined lateral migration as MP >40%^{109,110}, with MP \leq 40% as the reference category. Passive ROM for hip and knee extension was measured using a goniometer in a standardized position, and the value for the worst side was used for all analyses (www.cpunp.se). Data were dichotomized into either full hip or knee extension versus limited hip or knee extension (-5° or less), and the former was used as the reference category.

Statistics

For Studies I, III and IV, IBM SPSS Statistics 24 (IBM Statistics Inc., Armonk, NY) was used for the statistical analyses. For Study II, the analyses were performed using Stata (IC v.13, StataCorp LP). For Study IV, R was used to perform validation of the data¹¹¹. For all studies, the significance level was set to <0.05

Study I

Construct validity was evaluated for known groups based on GMFCS I–V by linear regression analysis (chi-square test for trend). Arithmetic average values was used for domain and total scores.

Test–retest reliability was calculated by intraclass correlation coefficients (ICC (A,1)) with two-way random, absolute agreement for single measures with 95% confidence intervals (CIs)¹¹². ICCs were calculated for each domain score and the total score, ranging from 0 to 100. The ICC should exceed 0.75¹¹³.

Internal consistency was analysed with Cronbach’s alpha, which shows the average of the correlations among all items in the measure, compared with the total score. To indicate relevant internal consistency, the value should exceed 0.8 for basic research and 0.9 for clinical instruments¹¹⁴.

Study II

The chi-square test was used to analyse differences between males and females; period prevalence was calculated by comparing children with scoliosis to the total population during the study period.

Kaplan–Meier analysis was used to identify the age at diagnosis of a) moderate or severe scoliosis and b) Cobb angle $\geq 40^\circ$. Both groups were stratified by age and GMFCS level. The values at risk are presented in 5-year intervals.

Cox regression analysis was used to compare the risk for scoliosis between different age groups, GMFCS levels, and in males and females. The model fulfilled the proportional hazards assumption.

Study III

For descriptive statistics, categorical data were reported as frequencies and percentages (n (%)) while discrete and continuous data were reported as median, means and standard deviations (SD).

The non-parametric tests chi-square and chi-square for trends (the linear-by-linear association test) were used to analyse the differences between variables.

Study IV

The variables for GMFCS level, sex, spastic CP, epilepsy, hip surgery, MP, passive hip extension and knee extension were analysed using logistic regression, giving odds ratios (ORs) with 95% CI for scoliosis. First, each variable was analysed using univariable logistic regression. Then, multivariable logistic regression analysis was used, removing one explanatory variable at a time (i.e., the variable with the highest non-significant p-value), in a stepwise backward elimination process. This was carried out until only significant variables remained in the model.

A risk score was constructed using the remaining variables as independent significant predictors of scoliosis. The risk score was evaluated using the area under the receiver operating characteristics (ROC) curve (AUC). The AUC can be interpreted as the probability that a randomly selected child with scoliosis has a higher predicted risk of severe scoliosis before the age of 16 than a randomly chosen individual without scoliosis. An AUC value of 1 is considered perfect and a value of 0.5 no better than chance¹¹⁵. To additionally validate the risk score AUC, a 10-fold cross validation was used. The risk score development process was also validated with a different predictor selection approach using L_1 -penalized logistic regression^{116,117}.

Ethics

Study I was approved by the Medical Research Ethics Committees at Uppsala University (2013-493) in Sweden and by the Regional Ethics Committee South-East (2013/1096) in Norway. Studies II–IV were approved by the Medical Research Ethics Committee at Lund University Sweden (LU-433-99, 383/2007), and permission was obtained to extract data from CPUP.

Results

Study I: The CPCHILD

Of the families included, approximately 60% had a girl. Most caregivers were the biological parents with a mean age of 40 years (range, 26–58). Mainly mothers (81%) answered the questionnaire; in 63 of the 64 families, the same caregiver answered both the first and second questionnaire. The average time to complete the questionnaire was 16 minutes. The families' mean total score for the CPCHILD was 63.1 ± 19.1 (range, 11.9–97.7); for total score and domain scores per GMFCS level see Table 2. These analyses were based on the first questionnaire for the 106 families with $\geq 80\%$ responses per domain (Figure 11).

Table 2.

CPCHILD scores according to GMFCS level. CPCHILD scores per domain and total, presented as mean and standard deviation for each GMFCS level.

	GMFCS level				
	I (n=21)	II (n=25)	III (n=21)	IV (n=17)	V (n=22)
Total score	83.8 ± 9.1	70.7 ± 13.4	64.3 ± 14.7	51.0 ± 12.6	43.0 ± 13.5
Domain score					
1 ADL, personal care	81.3 ± 16.7	55.2 ± 23.4	46.2 ± 21.9	28.1 ± 19.9	28.1 ± 19.0
2 Positioning, transferring	92.8 ± 9.17	75.4 ± 15.4	52.0 ± 19.0	31.0 ± 16.3	28.7 ± 14.7
3 Comfort and emotions	86.1 ± 12.7	85.8 ± 11.5	84.6 ± 12.5	78.9 ± 15.9	71.3 ± 21.4
4. Communication	80.8 ± 15.8	65.4 ± 19.0	72.2 ± 22.5	58.2 ± 21.1	34.4 ± 11.8
5. Health	70.1 ± 16.4	70.7 ± 14.8	69.7 ± 13.8	68.6 ± 13.3	60.3 ± 17.3
6. Quality of life	76.2 ± 16.3	75.2 ± 13.3	69.5 ± 17.5	70.6 ± 18.9	53.6 ± 25.7

The CPCHILD questionnaire showed construct validity and known-group validity ($p < 0.001$), with the ability to discriminate between GMFCS levels (Figure 12).

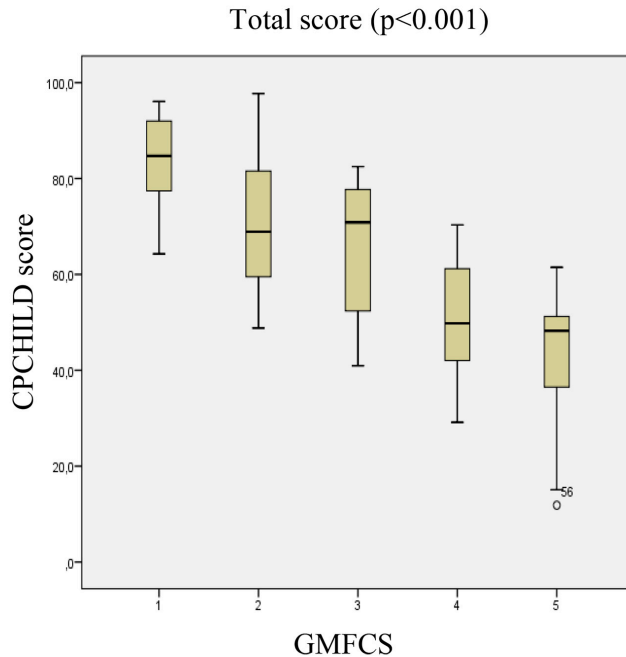


Figure 12. Construct validity with standardized total score for the CPCHILD compared with the known groups defined as GMFCS levels I–V; p-values were calculated using the linear regression analysis for arithmetic average values for the total score.

ICCs showed high test–retest reliability for the total score (ICC 0.92; 95% CI 0.88–0.95) and the domain scores (ICCs 0.72–0.92). There was also high internal consistency as measured by Cronbach’s alpha, with a total score of 0.96, and 0.83–0.96 for the domain scores. Both test–retest reliability and internal consistency were based on the 64 families who answered both surveys within the time limit of 4 weeks and with $\geq 80\%$ responses per domain (Figure 11).

Study II: Scoliosis

At the latest clinical examination, 15% (140/962) of the individuals had developed scoliosis, with 48 being graded as moderate and 92 as severe. Spinal fusion was performed in 54% of the individuals with severe scoliosis at a mean age of 14.1 years (range, 6–22). Slightly more of the females underwent surgery (57%), than of the males (52%) ($p=0.62$). The individuals who had surgery had GMFCS level III (n=2), IV (n=15) and V (n=53).

Radiographic examination was reported for 91% (128/140) of the individuals with moderate or severe scoliosis. The Cobb angle was $<20^\circ$ in 27 cases, $20\text{--}39^\circ$ in 14 cases and $\geq 40^\circ$ in 87 cases.

Kaplan–Meier survival estimations for clinical examination showed that the incidence of scoliosis increased with age and GMFCS level, and scoliosis was seen in younger ages in children with lower gross motor function (Table 3 and Figure 13). The analyses based on radiographic examination showed a similar pattern (Figure 14).

Table 3. Incidence of scoliosis according to clinical and radiographic examination at 10 and 20 years of age related to GMFCS level.

	GMFCS I-II	GMFCS III	GMFCS IV	GMFCS V
CE, age 10	1%	5%	10%	30%
CE, age 20	5%	30%	45%	80%
RE $>40^\circ$, age 10		2%	5%	20%
RE $>40^\circ$, age 20		8%	35%	75%

CE, clinical examination (moderate or severe); RE, radiographic examination (Cobb angle $>40^\circ$).

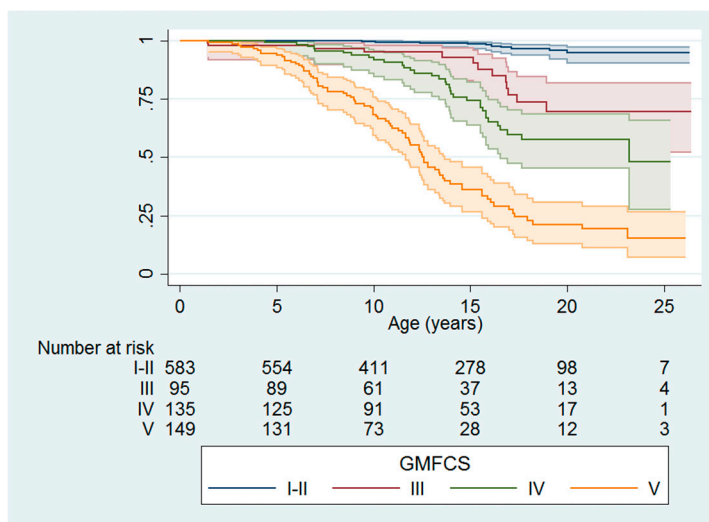


Figure 13. Survival function with 95% CI showing the risk of having a moderate or severe scoliosis diagnosed at different GMFCS levels and ages. Numbers at risk at inclusion and at 5-year intervals are reported.

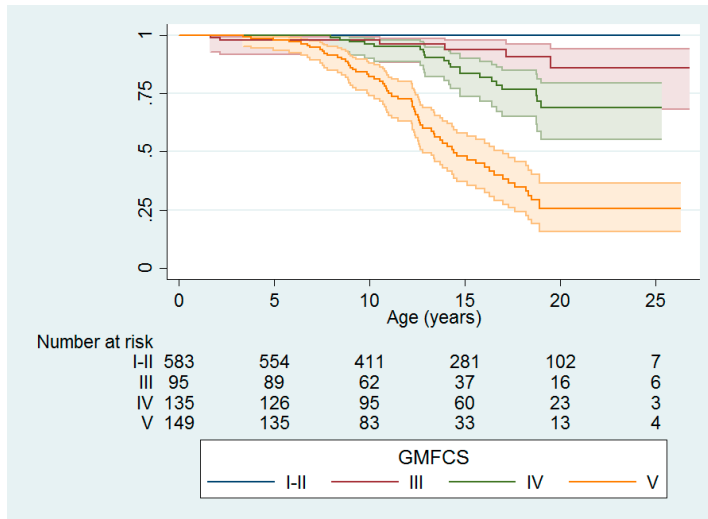


Figure 14. Survival function with 95% CI showing the risk of having a scoliosis with Cobb angle $\geq 40^\circ$ diagnosed at different GMFCS levels and ages. Numbers at risk at inclusion and at 5-year intervals are reported.

No child at GMFCS levels I and II developed scoliosis with a Cobb angle $> 40^\circ$. There were no sex differences based on the survival estimation; however, the hazard ratio (HR) was 1.4 for females versus males. A high GMFCS level indicated a high risk for scoliosis.

Study III: Spinal orthoses

Spinal orthoses were used by 9% (251/2800) of the children. The children's median age was 8.0 years, with a boy/girl ratio of 54/46. No child at GMFCS levels I and II used spinal orthoses. Spinal orthoses were more frequently used by children with higher GMFCS levels ($p < 0.001$), and the proportion of children using spinal orthoses increased with age ($p < 0.001$). Of those using spinal orthoses, 59% also had scoliosis. The remaining 41% without scoliosis used the spinal orthoses either to improve function or to prevent deformity.

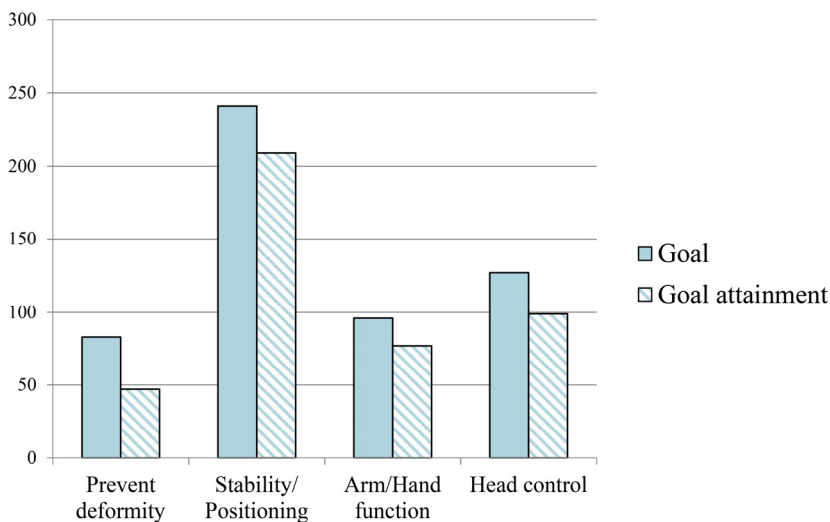


Figure 15. Number of children with each treatment goal and the rate of goal attainment.

The primary goal was to improve function for almost all children who used spinal orthoses including one or more of the following goals: improve stability/positioning (96%); improve arm–hand function (38%); and/or improve head control (51%) (Figure 15). Most children reported more than one goal. No significant differences were seen in either treatment goals or goal attainment levels in relation to age or sex.

Most children with low gross motor function (GMFCS IV and V) used spinal orthoses for stability/positioning (96%), compared with 75% of the children at GMFCS level III ($p=0.043$). The second most common goal was head control, chosen by slightly more children with severe scoliosis and low functional ability. Improved arm–hand function was the third most common treatment goal. Significantly more children at GMFCS IV (66%) compared with children with level III (25%) and level V (26%) chose this goal ($p<0.001$). The least selected goal was to prevent deformity (33%), with no significant differences in goal or goal attainment in relation to age or sex. The distribution per GMFCS level increased from 25% in GMFCS III to 35% for GMFCS V. For those with a more severe scoliosis at clinical examination, a higher proportion of children used spinal orthoses to prevent deformity.

Goal attainment was high for all four goals, from 87% for the goal of stability/positioning to 57% for the goal of preventing deformity (Figure 15).

Of the 2800 children, 17% (n=486) had scoliosis (information was missing for 18 children). Most children with scoliosis (70%) did not use spinal orthoses.

Scoliosis occurred as frequently in boys (17%) as in girls (17%), but a significantly higher proportion of girls (6.4%) than boys (3.7%) had a moderate or severe scoliosis ($p=0.002$), and twice as many girls (2.2%) as boys (1%) had a spinal fusion.

Study IV: Risk score for severe scoliosis

There were 14.1% (92/654) children with severe scoliosis before the age of 16, with 45.7% (42) males and 54.4% (50) females; 58.7% were classified as GMFCS V. Of the group of 92 children, 59 had undergone spinal fusion for scoliosis, six had a Cobb angle of $\geq 40^\circ$ and 27 had severe scoliosis identified at clinical examination.

Female sex, GMFCS levels IV and V, epilepsy and limited knee extension were all identified as significant predictors of developing severe scoliosis before the age of 16. This association determines the equation for calculating the risk score as:

Risk score = $-3.859 + 0.5 \cdot \text{sex} + 2.290 \cdot \text{GMFCS}_{\text{IV}} + 3.325 \cdot \text{GMFCS}_{\text{V}} + 0.586 \cdot \text{epilepsy} + 0.619 \cdot \text{knee extension}$.

Sex is a dichotomous indicator variable that took a value of 1 for female and 0 for male. GMFCS IV is also a dichotomous indicator variable that took a value of 1 when the individual had GMFCS level IV, and 0 otherwise. GMFCS V is a corresponding indicator. Epilepsy took a value of 1 when epilepsy was present and 0 when not. Limited knee extension corresponded to the value on the worse side when both sides were measured and took the value of 1 when the limited knee extension was -5° or less, and 0 when the individual had full knee extension. The risk score can be translated into the risk of developing scoliosis using Table 4.

Table 4.

The risk score for scoliosis translated into a risk score level, corresponding to the risk of developing severe scoliosis before the age of 16.

Risk Score	Risk of Scoliosis (%)
< (2.20)	0 to 10
(2.2) to (-1.39)	10 to 20
(-1.39) to (-0.85)	20 to 30
(-0.85) to (-0.41)	30 to 40
(-0.41) to 0	40 to 50
0 to 0.41	50 to 60
0.41 to 0.85	60 to 70
0.85 to 1.39	70 to 80
1.39 to 2.20	80 to 90
≥ 2.20	90 to 100

The sensitivity and (1 – specificity) of the risk score are shown in Figure 16. The discriminatory accuracy of the risk score was high, with an AUC of 0.874 (95% CI 0.836–0.913), indicating a strong ability to differentiate between high- and low-risk individuals. The AUC was marginally worse after cross validation (AUC=0.848).

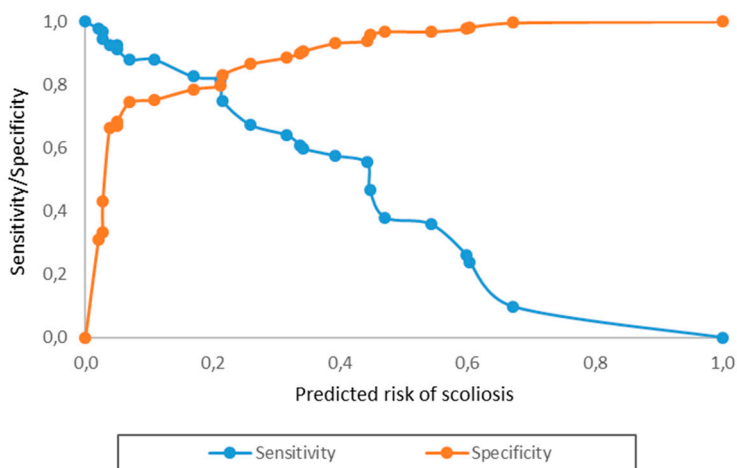


Figure 16. The proportion of children with scoliosis correctly predicted to develop severe scoliosis before the age of 16 (sensitivity) and the proportion of children without scoliosis correctly predicted not to develop severe scoliosis before the age of 16 (the specificity) for the choice of a cut-off to indicate a high-risk individual.

Discussion

The overall aims of this thesis were to enhance knowledge on how scoliosis develops in individuals with CP (Study II), to explore treatment goals and goal attainment with spinal orthoses (Study III), to differentiate children at high risk from those with low risk for scoliosis (Study IV) and to evaluate a questionnaire to assess HRQoL in children with CP (Study I).

We found an overall incidence of 15% for moderate or severe scoliosis in young adults with CP. Scoliosis is strongly correlated with GMFCS level; it starts early and continues into adulthood for those with severe motor impairment (Study II). GMFCS levels IV and V, female sex, epilepsy and limited knee extension are predictors for scoliosis and were used to develop a risk score to discriminate between high- and low-risk individuals (Study IV). High goal attainment and functional benefits of spinal orthoses in children with CP are reported (Study III). The CPCHILD is a sound proxy-reported instrument to measure HRQoL in children with CP in Scandinavia (Study I).

As a classification of gross motor function, the GMFCS provides the means to discriminate between groups of individuals with CP and to pinpoint those groups with greater difficulties depending on the problem examined¹⁹. All four studies confirmed distinct differences between the five GMFCS levels. In Study I, construct validity was high, showing that the CPCHILD questionnaire was able to discriminate between GMFCS levels. Studies II–IV showed a higher risk for scoliosis in individuals at GMFCS levels IV and V, while Study III revealed that only children in GMFCS levels III–V used spinal orthoses. To detect these differences, the entire spectrum of individuals from GMFCS I to GMFCS V need to be included. For early detection of scoliosis, it has been emphasized that follow-up programmes should be based on the child's GMFCS level, bearing in mind the child's age^{30,31,63}.

In Study II, the incidence of moderate and severe scoliosis was the outcome of interest. In Study IV, when differentiating between high- and low-risk individuals, only severe scoliosis was treated as outcome. The decision not to include children with mild scoliosis in these studies was based on psychometric evaluation of the clinical examination in CPUP, which indicated that most children with mild scoliosis only had a Cobb angle of 5–15°³². In Study IV, only severe scoliosis with a Cobb angle of $\geq 40^\circ$ was used, because it has been found to predict significant progression of the magnitude of the curve^{64,57} and is suggested as a

cut-off when considering surgical interventions ^{57,30,31}. However, at present, there are no internationally agreed criteria for the recommendation of spinal surgery ⁷⁶. In the analyses of spinal orthoses in Study III, all degrees of scoliosis (mild, moderate and severe) and also children without scoliosis were included and described.

Scoliosis in a total population

A neuromuscular scoliosis is usually progressive, even after spinal growth is completed ³¹, in contrast to idiopathic scoliosis where the risk of further progression is much lower ³⁵. Increased knowledge of the incidence of scoliosis in an unselected group of people with CP may contribute to guidance for predicting future risk for scoliosis, identify critical ages for surveillance, and ultimately, support the creation of guidelines for treatment.

Persson-Bunke et al. analysed the incidence of scoliosis in 666 children aged 0–18 years in southern Sweden based on data from 1995 to 2008. Because no relationship between CP subtype and scoliosis was found in that study, subtype was not included in Study II ³⁰.

The aim of Study II was to further analyse the incidence and prevalence of scoliosis related to gross motor function and age, but also in relation to sex, which was not investigated earlier ³⁰. Data were extracted from the same area and the cohort was expanded with longitudinal data between 2008 and 2016. A further 296 individuals were included and the age was increased up to 25 years.

The main findings in Study II were that higher GMFCS level was a significant risk factor for the development of scoliosis, that scoliosis occurred at younger ages in individuals classified at higher GMFCS levels, and that the incidence of scoliosis continued to increase up to the age of 20–25 years.

Before the development of the widely used GMFCS levels, to be non-ambulatory was considered a risk factor for the development of scoliosis and the incidence of scoliosis directly paralleled the severity of the neurological impairment ^{41,57}. For children at GMFCS levels I and II, there is a low risk of developing neuromuscular scoliosis ³⁰, similar to that of adolescent idiopathic scoliosis in typically developed children ⁶².

The larger cohort and longer follow-up time in Study II made the separation of GMFCS levels IV and V possible, in contrast to Persson-Bunke et al. ³⁰. A higher GMFCS level is a significant risk factor for the development of scoliosis, as it is strongly associated with the child's GMFCS level. When examining the HR for developing clinically moderate or severe scoliosis in relation to GMFCS level,

GMFCS IV was shown to have an HR of 15, while it increased to 53 in GMFCS level V, compared with GMFCS levels I and II. It was shown that GMFCS level was the strongest, statistically significant independent risk factor for scoliosis³⁰, also reported by other groups^{118,84}. Bertoncelli et al. found no correlation between scoliosis and GMFCS level. However, in their study, they only included children with scoliosis at GMFCS levels II–V, and compared those with mild versus severe scoliosis; in addition, their findings were based on a small sample size, especially for GMFCS levels II and III^{56,66}. In contrast, in Study II, we included children with and without scoliosis and had larger samples at all GMFCS levels.

The reported incidence of scoliosis in people with CP varies^{59,31}. It is difficult to draw comparisons with other studies because of different definitions of scoliosis or different selected groups of individuals^{41,58,57}. Persson-Bunke et al. found that the overall prevalence of scoliosis was 29% in their initial cohort, including all degrees of scoliosis. However, when they only included moderate and severe scoliosis, the prevalence was 11%³⁰. In Study II, 15% developed moderate or severe scoliosis during the follow-up period. This difference may be due to the longer follow-up time and older ages of individuals that were included in Study II.

One of our main findings was that scoliosis occurred at younger ages in individuals with higher GMFCS levels (lower motor function), that is, already at 5 years of age for children in GMFCS V, while others have reported that children in most cases were diagnosed after 8 years and up to 13 years of age^{30,56}. In their study, Saito et al. included Japanese institutionalized residents and reported that 88% of children with radiographs taken before 10 years of age had developed scoliosis⁵⁷. In 1992, Terjesen et al. proposed that individuals with low gross motor function and whole body involvement should be followed with clinical examinations and radiographs from around 5 years of age⁷⁴. Children who are included and who participate in CPUP are regularly followed with clinical and radiographic examinations²⁷.

Curve progression occurs even after completion of growth in adolescence^{57,118}. Study II found that the incidence of scoliosis continues to increase up to 20–25 years of age, and at the age of 20, 75% of those at GMFCS level V had a Cobb angle $\geq 40^\circ$. This accords with Saito et al. who found the progression of scoliosis to continue beyond the age of 20. In 25% of individuals, progression stopped at the mean age of 22 years, but continued to progress in 75% of the cases⁵⁷. Others have shown that the mean Cobb angle increases with age up to 30 years¹¹⁸. Oda et al. only included children at GMFCS levels IV and V, and observed that the peak incidence of scoliosis occurred in the group aged 20–29 years, and that the Cobb angle increased more in the 20–29 age group than in the 10–19 age group¹¹⁸. This has clinical implications, namely the need for close surveillance into adulthood especially of those with low gross motor function.

Not surprisingly, the overall prevalence of CP was almost identical in Studies II–IV and the distributions were consistent with the overall boy/girl ratio reported for CP, that is, slightly more towards boys⁹. The incidence for boys is higher compared with girls with an overall boy/girl ratio of 1.4:1⁹. For secondary conditions like scoliosis³¹, pain^{119,120}, back pain⁶⁹, or even mental health or peer problems when having recurrent muscular pain^{121,122}, evidence for sex differences is increasing with girls being more afflicted, in contrast to comorbidities in CP, where no sex differences are found¹³.

In this thesis, we found sex discrepancies regarding scoliosis in all studies analysing scoliosis. A higher proportion of females had moderate or severe scoliosis in Studies II and III, compared with males who more often had mild scoliosis. In Study II, the HR was evaluated for the development of moderate or severe scoliosis in relation to sex, giving an HR of 1.4 for females versus males. This means that the relative incidence is 40% higher for girls to develop moderate or severe scoliosis than for males. For spinal surgery, the same pattern was detected, with girls more often having spinal surgery. To our knowledge, this trend in sex differences for moderate/severe scoliosis and spinal surgery seen in Studies II and III has not previously been reported for children with CP. In Study IV, female sex was shown to be a predictor for severe scoliosis, as also recently reported elsewhere^{56,66}. Bertonecchi et al. found female sex to be a risk factor for severe scoliosis⁵⁶, and this was later confirmed when validating their clinical prediction model⁶⁶.

The connection between the spine and the pelvis gives rise to multifaceted questions and decisions when discussing appropriate and adequate interventions. Scoliosis is sometimes preceded by hip displacement/dislocation or windswept hip deformity, which can cause a pelvic obliquity and initiate scoliosis^{123,124}. In non-ambulatory individuals, hip dislocation occurs first, followed by pelvic obliquity and then scoliosis in 75% of cases¹²³. Ágústsson et al. showed higher odds for both scoliosis and windswept hips for adults with CP who were immobile in a lying position for long periods (>8 hours) and who were unable to change position compared with those who were able to move and change position; lying solely in a supine position resulted in higher odds of windswept hips.

In supine lying, limited knee or hip extension might cause the legs to tilt over to one side. This deviation may be increased over time and with gravity, and ultimately becomes a severe windswept deformity, thereby affecting the spine. Even a small unilateral limited knee extension creates an apparent leg length discrepancy, which in standing and walking leads to an oblique pelvis, if not compensated for by toe walking, and compensatory postural curve of the spine that could become structural over time⁶⁷. Both scoliosis and hip dislocations are associated with postural asymmetries in lying, sitting and standing¹².

Our findings in Study II provide increased knowledge of scoliosis and its incidence in an unselected group of people with CP. This can contribute to identifying the critical ages for surveillance and guidelines for treatment. Surveillance programmes should be based on age and GMFCS level and must start at young ages and continue into adulthood.

Prediction of scoliosis

Early identification of children at high risk for scoliosis is crucial to optimize follow-up programmes and to allow preventive strategies from an early stage, and thereby hopefully reduce the incidence of severe scoliosis in children with CP. This requires the ability to discriminate between high- and low-risk individuals.

The aim of Study IV was to predict the risk for development of severe scoliosis before the age of 16 in children with CP, and to discriminate between high- and low-risk individuals at the age of 5 based on the following potential risk factors: GMFCS level, sex, spastic CP, epilepsy, hip surgery, MP, passive hip extension, and knee extension.

Study II confirms earlier findings that scoliosis can start early^{57,30,118,31}, but also progresses rapidly after onset⁸⁴, which clearly reinforces the importance of early identification. While most children with CP are born without any deformities, postural asymmetries appear even in young children with CP, and a preferred lying posture during the first 12 months of life can give rise to an asymmetrical posture that over time becomes established as tissues adapt^{90,79}. The Hueter–Volkman law⁴⁸ helps us understand why an asymmetrical posture can cause deformities such as scoliosis. A vicious cycle is created, initiated by a prolonged asymmetric loading of the vertebrae (e.g., poor lying, sitting and standing positions), which can cause asymmetric growth of vertebrae. This in turn leads to wedging of the vertebrae, thereby creating a scoliosis⁴⁶ that further affects the posture and increases the need for proper positioning. Spasticity can also cause functional limitations and postural asymmetries in children with CP. In a total population, spasticity seems to increase in most children during their first years, peaking at 5 years of age, and thereafter decreasing. Children with higher GMFCS levels and lower functional levels also had a higher intercept (a higher muscle tone at the starting point of the model), and had a more pronounced rate of increase and decrease of spasticity levels^{125,126}. The most important and rapid growth spurt in children generally occurs from 11 to 14 years of age³⁵, when scoliosis tends to develop more rapidly. The risk score was therefore developed based on risk factors identified at the age of 5 to predict severe scoliosis within 10 years.

A variety of potential risk factors for scoliosis have been suggested in the literature: GMFCS level ^{30,63,31}, age at onset ^{118,31,30,57}, magnitude of the curve ^{57,60,30,31}, pelvic obliquity ¹²⁷, windswept hips ^{124,67}, hip dislocation ^{70,71} and positioning ^{79,12,67}. Bertonecchi et al. examined the risk factors for severe scoliosis ⁵⁶. Their univariable analyses revealed the following risk factors for scoliosis: history of previous hip surgery, intractable epilepsy and female sex ⁵⁶, which were later confirmed as predictors for severe scoliosis through a validation study ⁶⁶. Study IV included both univariable and multiple regression analyses. In contrast, Bertonecchi et al. reported their analyses in two separate articles, with unilateral logistic regression analyses in one ⁵⁶ and multiple regression analyses in the other ⁶⁶. Study IV could confirm two of the variables suggested by Bertonecchi et al., namely female sex and epilepsy, but not previous hip surgery. This variable was found to be significant in the univariable analyses but non-significant in the multivariable analyses when controlling for other variables. The reason for this probably lies in the different statistical approaches adopted in the two studies. Differences in study design could also have contributed to the different outcomes, with the cross-sectional design used by Bertonecchi et al., as against our prospective design with longitudinal data. Moreover, the population sample differed. Bertonecchi et al. included 120 individuals aged 12–18, while Study IV included 654 individuals aged 5 and 16 years of age.

Most of the children included in Study IV have been followed regularly and received early interventions such as hip surgery in accordance with the CPUP protocol and guidelines. Thus, by using a preventive follow-up programme (that might include early corrective interventions), protective effects can be given for both hips and spines ^{27,128}. This way of implementing early interventions may differ from other countries without follow-up programmes where hip surgery has been suggested as a risk factor for scoliosis ^{56,66}.

The statistical model used for calculating the explanatory variables and creating the risk score in Study IV has been described by Hermanson et al. ¹⁰⁹. They created a risk score for hip displacement in children with CP that included GMFCS level, age, initial MP and head-shaft angle. The discriminatory accuracy of the CPUP hip score was high (AUC=0.87), indicating a strong ability to differentiate between high- and low-risk individuals for hip displacement. This was similar to the result in Study IV, even though the explanatory variables and the outcome differed. Both risk scores use cut-offs for hips and spines that are likely to require surgical treatment to prevent further progression into hip dislocation and collapsing spine.

Our risk score for scoliosis was based on female sex, GMFCS levels IV and V, epilepsy, and having limited knee extension, which were all identified as independent predictors for the development of scoliosis before the age of 16. The

AUC of the resulting risk score was 0.874, and like Hermanson et al.¹⁰⁹, the result indicates a high accuracy in differentiating between high- and low-risk individuals. The AUC remained at this level after cross validation, showing that its high value was not due to overfitting, and thus may be generalized to other populations.

Our results are consistent with previous findings^{31,30} in identifying GMFCS level as a strong predictor of scoliosis, with an OR of 9.86 for GMFCS level IV up to 28.96 for GMFCS level V. The OR is a measure of association between exposure and an outcome, by comparing the relative odds of the occurrence of the outcome of interest (e.g., scoliosis) given exposure to the variable of interest (e.g., CP)¹²⁹. When only including the variable of GMFCS level in the ROC analysis, the AUC remained high at 0.85. When including all GMFCS levels, the ROC analysis reflected the diversity in individuals with CP at different GMFCS levels. The GMFCS is the basic core when predicting and planning interventions in children with scoliosis.

Epilepsy is a common co-causal comorbidity in CP^{130-132,13}. Individuals with quadriplegia (GMFCS level V) are reported to have epilepsy more often and to have an earlier onset than individuals with diplegia or hemiplegia^{133,134}. Bertonecchi et al. analysed epilepsy as a potential risk factor for scoliosis and divided the condition into three groups: no epilepsy, well-controlled epilepsy and intractable epilepsy. Their study indicated that patients with intractable epilepsy were significantly more likely to develop severe scoliosis, in contrast to those without intractable epilepsy and those with well-controlled epilepsy⁵⁶. Study IV confirmed these findings, with epilepsy being a predictor of scoliosis even after adjustment for other variables.

Limited hip and knee extension is highly associated with postural asymmetries¹², with knee contractures being the most common^{12,67,94,78,135,136}, especially for those with a low level of motor functioning^{94,78}. Ágústsson et al. showed that adults with knee contractures had higher odds of scoliosis and windswept hips⁶⁷. Therefore, in Study IV, it is not surprising that limited knee extension contributed to the risk of developing moderate or severe scoliosis. Cloodt et al. also showed that limited knee extension exists at all GMFCS levels and increases with age, and is present in almost 25% of children up to 15 years of age⁹⁴; equally common in adults¹². Brantmark et al. found that the decline in gross motor function in adolescents correlates with limited passive joint range of motion and that those using a wheel-chair for mobility had the greatest deficits in knee extension⁷⁸.

Postural asymmetries may cause progressive deformities, making it difficult to find comfortable positions in both lying and sitting¹². Preferred positions may over time become habitual and cause further deformities⁶⁷, thereby creating a vicious cycle that is difficult to stop. Limited hip and knee extension may co-occur⁶⁷, which might explain why limited hip extension did not remain significant in the multivariate regression analyses.

Limited knee extension also affects standing posture¹² and can lead to deviating gait patterns⁷⁸, causing knee pain¹³⁷ that increases with age¹¹⁹. Musculoskeletal pain is most common in the lower extremities¹¹⁹ and is also associated with decreased joint mobility¹³⁸. Limited knee extension also seems to decrease walking ability in individuals in GMFCS levels I–III¹³⁸. Continuing to be an ambulator into adulthood demands successful prevention of knee contractures⁷⁸. It is vital to monitor ROM and posture from an early age and to promptly address contractures and postural asymmetries. The association between different deformities and postural asymmetries shows the value of hip surveillance programmes in reducing the frequency²⁶.

The ROC curve (Figure 16) shows consecutive cut-offs for the predicted risk for scoliosis, and shows the possible cut-offs for classifying individuals as high or low risk¹¹⁵. For a clinician to make a reasonable assessment of the risk for scoliosis, it would be advisable to base the decision about cut-offs on the nature of the treatment option being considered¹⁰⁹. For minor interventions like spinal orthosis, one should try to capture as many children as possible so as not to risk missing someone (high sensitivity). We analysed the same risk factors to predict moderate scoliosis and a Cobb angle $>20^\circ$ before the age of 16 and the outcome was similar to that found for severe scoliosis. However, for major interventions like surgery, it is important to select a more conservative cut-off that only includes high-risk individuals to prevent unnecessary interventions that in turn can have a negative impact on QoL (high specificity). In general, spinal surgery is not performed before severe scoliosis is already present. However, our risk score may identify children who need close clinical and radiographic surveillance of their spine.

In Study IV, we created a risk score that showed a high AUC and discriminatory accuracy for differentiating between children with CP. It will hopefully provide clinicians with early insight regarding a child's risk for developing severe scoliosis, allowing them to differentiate between high- and low-risk individuals with scoliosis and CP. To our knowledge, this is the first study to create a risk score for development of severe scoliosis based on predictors identified in 5-year-old children with CP.

Functional benefits of spinal orthoses

Appropriate postural support is vital for those unable to maintain or change position, or having difficulties with mobility^{67,12,78,79}. In addition, with scoliosis being a lifelong condition in many cases^{57,60,31}, the functional benefits with spinal orthoses^{77,80,74,73,81} are important considerations for health care services to optimize QoL and participation for individuals with CP.

The aim of Study III was to analyse the treatment goals and goal attainment with the use of spinal orthoses in a population of children with CP, and to describe the use of spinal orthoses in relation to age, sex, gross motor function and scoliosis. To our knowledge, this is the first study to describe the use of spinal orthoses in a total population of children with CP.

Due to the different study designs and populations included in different studies, it is difficult to make comparisons of the overall prevalence of spinal orthoses. Studies of orthopaedic interventions seldom report the use and frequency of spinal orthoses; on the other hand, some studies of spinal orthoses only include participants that use one. In the literature, the number of reports preferring surgical correction is far higher^{77,85,139-141} than the number supporting the non-invasive method of spinal orthoses^{81,74,73}, making recommendations about spinal orthoses and evaluation of their potential benefits a rather controversial topic⁸⁴.

Study III showed that spinal orthoses seem to have functional benefits, with a high overall rate of goal attainment. Almost all children used spinal orthoses to improve function with one or more of the following goals: to improve stability/positioning; to improve arm–hand function; or to improve head control.

Nine percent of the children aged 1–14 years with CP used a spinal orthosis. Numerous goals were reported for most children: 547 goals were reported for the 251 children who used spinal orthoses. Children in GMFCS level IV reported most goals per child (a mean of three goals/child), followed by level V (two goals/child) and III (one goal/child), reflecting the different functional needs between the three GMFCS levels. The use of orthoses increased with age, from <4% in 2-year-olds to 13% in 9-year-olds; spinal orthoses were equally common regardless of sex, despite sex differences earlier described for scoliosis. There were no significant differences in either treatment goals or goal attainment levels in relation to age or sex. The increased use with age may partly be explained by the fact that scoliosis also increases with age^{31,30}. It may also reflect the increased demands of independence for individuals with CP in all environments, especially at school and in the community, given that a large proportion of the children did not have scoliosis, but used spinal orthoses mainly for functional reasons.

As stated in the definition of CP, children and adults have difficulties with movement and posture¹, which in turn can give rise to problems with stability^{73,55,58,142}. This may explain why spinal orthoses were only used by children at GMFCS levels III–V, that is, those with the lowest motor functioning. The use of spinal orthoses ranged from <2% at GMFCS level III to 38% of all children classified at GMFCS level V.

The overall primary goal for families and physiotherapists in Study III was stability/positioning, chosen by 96% of the individuals wearing a spinal orthosis,

and with an equally high goal attainment of 87%. These high percentages illustrate the great need that these children have to gain stability in their daily life, and concomitant high goal attainment indicates that spinal orthoses improve stability/positioning. In a study on soft spinal orthoses, Letts et al. found that stability/positioning was enhanced for 90% of the included participants⁸¹. In contrast, Terjesen et al. reported that truncal stability was not necessarily improved, especially not in children with long thoracolumbar curves⁷⁴. These differences in findings may be explained by the different populations studied; for example, Terjesen et al. included individuals with a mean Cobb angle of 93° (range, 40–145°).

Stability/positioning as a treatment goal was reported more frequently for children at GMFCS levels IV and V than for children at level III, who usually require less trunk support. More than 30% of children with CP are non-ambulant, and spend most of their lives in sitting or lying positions⁸. In a study by Kalen et al. on untreated scoliosis in adults with CP, >50% were non-ambulators⁷⁰; being a non-ambulator increases the risk for scoliosis^{67,12}. For those who are non-ambulant, the sitting position is vital and also a necessity for performance of vital tasks of daily life, such as transportation⁸⁹, with sitting clearly affecting the QoL in individuals with CP, as they spend long hours in this position^{84,81,77,85,83}.

Trunk stability may improve head control^{73,74}, and the treatment goal of head control was reported for 51% of the children, being the second most common goal; goal attainment was 78%, which accords with findings elsewhere⁸¹. This treatment goal was chosen slightly more often by those with severe scoliosis, with 70% of children in GMFCS level V. The high distribution of low motor functioning (i.e., higher GMFCS levels) and lower goal attainment for this goal compared with goal stability (87%) can perhaps be seen as an indication of the great difficulties children with severe postural asymmetries face. Head control can be challenging for children with lower levels of motor function but it is vital to optimize function and participation, and is closely connected to abilities such as communication and eating⁸⁹.

A lack of stability may affect arm–hand function and lead to reduced manual wheel–chair mobility⁹³, whereas improved arm–hand function can enhance play, work and independence in the activities of daily living^{74,73,81}. Improved arm–hand function was a treatment goal for 38% of the children, of whom 80% attained the goal. More than twice as many children at GMFCS level IV (51/77) used a spinal orthosis to improve arm–hand function compared with children with levels III (1/4) and V (44/170). This highlights the relationship between stability and arm–hand function, but perhaps also the necessity for this group of children to gain more independence and increase their mobility.

It has previously been reported that the main indication for the use of spinal orthoses in individuals with CP is to prevent, stabilize or delay the progression of scoliosis^{80,74}. Only one-third of the children in Study III used a spinal orthosis to prevent or correct a spinal deformity, making it the least common treatment goal; goal attainment was 57%, which is consistent with the finding of Olafsson et al.⁸⁰. Seven in 10 children with mild scoliosis, but fewer than four in 10 with severe scoliosis, attained the goal of preventing spinal deformity, which indicates that the rate of goal attainment was proportional to the severity of the curvature. This may be explained by the better outcomes reported for spinal orthoses when used for smaller curvatures with radiographic Cobb angles $\leq 40^\circ$ ^{80,74}, and perhaps also by the Hueter–Volkman law⁴⁵. The law states that when applying asymmetric compression forces on a vertebrae, wedging can develop due to a combination of asymmetrical growth, vertebral body remodelling, and epiphyseal wedging⁴⁸, thereby giving rise to the vicious cycle of scoliosis progression⁴⁶. Animal studies have shown that asymmetric growth, which may develop due to bad positioning, is more important in younger animals regarding the development of scoliosis⁴⁸. This knowledge further reinforces the fact that focus should be on preventive and early treatment aiming to maintain a neutral position of the spine especially in growing individuals^{84,80}, rather than on efforts to stabilize a collapsing spine at a later stage.

Of the 251 children who used spinal orthoses, 59% had scoliosis. Of the children at GMFCS level I, 0.3% had moderate scoliosis, while 6.4% at GMFCS level IV and 22% at GMFCS level V had moderate or severe scoliosis. No clear association was found between the severity of the scoliosis and goal attainment for any of the three functional goals.

A large proportion (41%) used spinal orthoses without having a scoliosis, either to improve function or to prevent deformity. This is a novel finding. Bertoncelli et al. found that patients with truncal tone disorders were more likely to develop severe scoliosis than those without⁵⁶, and spinal orthoses can be one way of managing a truncal tone disorder like hypotonia, and not necessarily a scoliosis. Olafsson et al. showed that the greatest success for spinal orthoses in individuals with scoliosis was seen in those having hypotonia and being ambulators⁸⁰. This achievement was perhaps due to small curves that were managed at an early stage of the scoliosis and to the fact that the individuals were mobile, indicating a better gross motor function.

One methodological consideration is that Sweden has a public health care system that generally functions well. We have free health care, and orthotics and assistive devices are available for children without any cost to the families. This may affect the use of spinal orthoses in both Studies II and III because the interventions given and the use of spinal orthoses bear no relation to the economic situation of the

family. However, this may clearly differ in countries where there are no surveillance programmes or free health care. Moreover, because there is no international consensus on the use of spinal orthoses, their use is likely to vary.

Although the goal of using spinal orthosis to prevent spinal deformity remains important, the functional benefits (i.e., stability, head control and arm–hand function) in daily life, appear to be of greater importance. Because the ultimate treatment goal is to improve activity, participation and QoL, children with postural deficits should be given the opportunity to explore the functional benefits of a spinal orthosis.

The CPCHILD: a sound proxy-reported questionnaire for HRQoL

A disease-specific questionnaire, the CPCHILD, was developed in Canada for children with the most severe forms of CP. It has been shown to be a valid and reliable measure of HRQoL¹⁰³ and has been validated in other countries^{104,106,105}. HRQoL focuses on the effects of illness, and specifically on the impact that treatment may have on QoL¹⁰⁰. It has been suggested that disease-specific instruments tend to be more sensitive to treatment-related changes¹⁰⁰, and generally more sensitive to the nuances of that particular condition¹⁴³.

The aim of Study I was to examine the test–retest reliability and construct validity of the Scandinavian caregiver version of the CPCHILD for use in children with CP.

Appropriate use of the CPCHILD includes following children’s HRQoL over time and measuring the effectiveness of interventions intended to improve or maintain HRQoL¹⁰³. With postural problems playing a central role in the lives of children with CP⁸⁹, with a higher incidence of pain and stiffness^{144,119,69}, from a life perspective, it is necessary to evaluate HRQoL for any treatments and interventions given. In a systematic review of measures of HRQoL, Carlon et al. noted that based on the collected psychometric and clinical utility data of the studies reviewed, the CPCHILD was one of the strongest outcome measures when evaluating HRQoL in school-aged children with CP¹⁴⁵.

The CPCHILD was created as a HRQoL instrument, but as such, it only includes a few items about the child’s health and QoL. The domains that address this topic contain just four items: *Overall quality of life* (1 item) and *Health* (3 items), with the following questions for *Health*: the number of visits to the doctor or hospital, the rating of overall health and the number of medications during the previous 2 weeks.

In contrast, the other functional domains contain as many as 24 items related to the child's health and QoL: *Activities of daily living/personal care* (9 items), *Positioning, transferring and mobility* (8 items) and *Communication and social interaction* (7 items). However, function is of central importance for individuals with CP, as the questionnaire is based on recommendations from caregivers, health care professionals and a review of other questionnaires¹⁰³. When analysing sitting (as a functional measure) in relation to HRQoL, Kolman et al. showed that increased comfort was associated with higher HRQoL⁸³, and that sitting balance for individuals with scoliosis affected caregiver satisfaction the most^{84,81,77,85}.

The initial hypothesis that children with lower motor ability (i.e., higher GMFCS levels) would have lower scores on the CPCHILD^{103,106,105,104} was confirmed in Study I. Known-group validity for the total and domain scores differed significantly according to the children's GMFCS level, as shown previously^{103,105,104,106}. We found smaller differences between GMFCS level for the domains *Comfort and emotions*, *Health* and *Overall quality of life*. The same has been shown for the German version of the CPCHILD for the domains *Comfort and emotions* and *Overall quality of life* when evaluated for children in GMFCS levels III–V, even though that study used other statistical methods¹⁰⁵. However, this was not seen in the original Canadian version or the Korean version, in which there was a significant difference between all domains^{106,103}. One possible explanation for this is that our findings indicate that more subjective domains such as *Overall quality of life* are less affected by function because the GMFCS levels are categories of only gross motor function. Rosenbaum et al. showed that for children with CP, functioning is only weakly related to the domain of *Overall quality of life*¹⁴⁶. The more functional domains *Activities of daily living/personal care*, *Positioning, transferring and mobility* and *Communication and social interaction* showed stronger correlations with GMFCS levels.

Not surprisingly, there was less difference between GMFCS levels for the *Health* domain. One possible interpretation for this is that people with disabilities can perceive their health as satisfactory despite having significant health problems, this is known as the disability paradox¹⁴⁷, and this may be especially true in relation to gross motor function¹⁴⁶. Thus, it is possible that a person with a lifelong disability will eventually normalize any difficulties, thereby internalizing the problem, and with time, their disability does not affect the individual's perceived HRQoL^{148,149}.

The test–retest reliability of the CPCHILD was substantially higher for the Scandinavian version (ICC total score of 0.92) compared with previous studies (ICC total score of 0.52–0.78)^{105,104,106}. However, no reliability coefficient as high as that of the original Canadian study have been reported (ICC total score 0.97)¹⁵⁰. The longer time limit of 4 weeks to return questionnaires did not affect the

reliability scores negatively, unlike in the case of the Korean version with the same time limit (ICC total score of 0.52).

Internal consistency was high for the Scandinavian version. Cronbach's alpha values of 0.83–0.96 exceeded the minimum of 0.8 for basic research for both the total score and all domain scores. However, only the total score and three of the six domains surpassed the threshold of 0.9, which is considered appropriate for a clinical instrument (Table 3) ¹¹⁴. Other CPCHILD versions have shown slightly lower internal consistency with alpha values of 0.81 to >0.90 ^{104,105}. Only the Korean version has shown an internal consistency as high as the Scandinavian CPCHILD. However, a high value can also indicate that the correlation between items is too high and some items may be redundant for assessing a single domain ^{113,151}.

The CPCHILD is available in two versions: one for caregivers and the other for children. We evaluated the version for caregivers. Initially, we also invited the children with CP to answer the child questionnaire, but because of the low response rate, analyses and comparisons could not be made. Thus, the effect of the proxy report should be considered when interpreting our findings.

Nowadays, it is recommended that, whenever possible, children should be asked about their perception of their QoL. However, difficulties may arise if the children in a study are very young or have disabilities that impede communication or cognition ⁹⁸. With such children, the use of proxy reports may be the only solution ¹⁰¹. Parents of children with a disability, as against children with no disability, are usually better able to estimate and report their child's functional difficulties, because they are more aware of them and must address their child's special needs ⁹⁸. Arnaud et al. found that the parent-reported QoL for children with CP was strongly associated with impairment ¹⁵². It is very important that the person who is closest to the child answer the questionnaire ¹⁵³; in our case, it was mostly answered by mothers (80%), which is likely a reflection of daily life for families with disabled children. One strength of the study is that the same caregiver answered both questionnaires in 63 of the 64 families. Research has shown that parental sex and relationship to the child might influence their HRQoL proxy report ¹⁵⁴.

Study I is a validation study, using the mean value for each domain as a way to investigate the validity and ability of the CPCHILD to discriminate between known groups and not as an evaluation of HRQoL per se. However, because all five validation studies report mean values, it is possible to compare proxy-reported HRQoL between countries for children with different GMFCS levels. The studies from Canada, Sweden and Korea included children for all five GMFCS levels ^{103,106,155}, while the German study included three levels (III–V) ¹⁰⁵ and the Dutch study included two (IV and V) ¹⁰⁴. The Korean study showed the largest difference

between the different GMFCS levels, ranging from 91.9 for level I to 24.3 for level V, a dramatic decline of ~67 points (Figure 17). While the findings from Sweden and Canada were very similar for each domain level, both countries showed a more even distribution of the different GMFCS levels. Thus, the GMFCS again shows its strong ability to differentiate between levels. The different results between countries indicate the similarities and differences in health care, and possibly cultural differences.

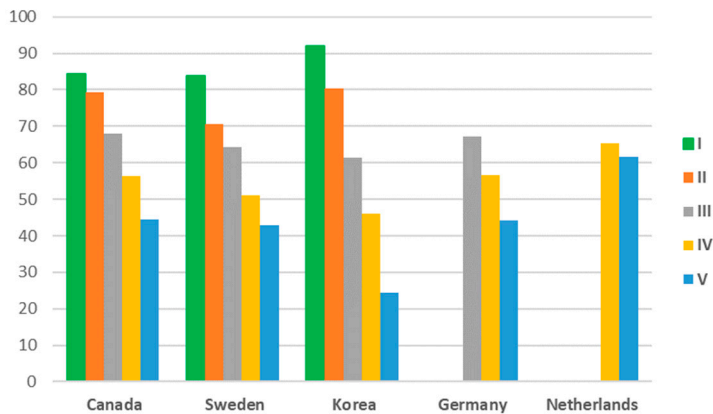


Figure 17. The five different validation studies and their means of the CPCHILD scores for the different GMFCS levels.

The identification of appropriate or inappropriate interventions is necessary to evaluate the impact they may have on QoL¹³. Comparisons of HRQoL foster discussion about interventions and health care for people with CP from the individual's perspective, making it possible to find good examples of the care given.

The Scandinavian version of the CPCHILD showed construct validity, high test-retest reliability and internal consistency for use in children with CP. Known-group validity for the total and domain scores of the Scandinavian version differed significantly according to GMFCS level. Thus, the CPCHILD appears to be a valid and reliable proxy-reported measure for HRQoL.

Limitations

Study I: The CPCHILD

For test–retest reliability, there were irregularities between two of the answers, which were reported as “not possible” at one occasion and as “no problem at all” at the other for children who required total assistance. They were included in the analyses because removing them would risk a false improvement of test–retest reliability. Few families chose to participate in the survey, which may be due to a questionnaire that was too extensive or because it was too difficult to interpret and answer. Another explanation may be that the difficult life situation many families live in (with many commitments and responsibilities) makes this questionnaire an additional task to complete. Most questionnaires were answered by the mothers, which may introduce a sex bias. The self-reported child version of the questionnaire was not validated.

Study II: Scoliosis

Not all individuals with moderate or severe scoliosis on clinical examination were examined radiographically. Some of the radiographic examinations were performed in the lying position, which might have underestimated the curve magnitude in children with a flexible curve. The numbers at risk in the Kaplan–Meier analysis were low at some GMFCS levels at 20–25 years of age.

Study III: Orthoses

The treatment goals and goal attainment levels were reported by the child’s physiotherapist, together with the child and the caregivers, based on their performance in everyday life and not in a clinical setting. Although this means that there was a subjective component to the assessment, it demonstrates how goal attainment was perceived by the families in various settings and conditions. The number of children wearing spinal orthoses differed between GMFCS levels, with only a small number at GMFCS level III. The study also lacked detailed information regarding the type of spinal orthoses, including their material and whether they were prefabricated or individually moulded. This was a cross-sectional study, where the design was used to document the status of a group at a particular point in time. It does not reflect changes over time.

Study IV: Risk score

By using data from children included in a preventive follow-up programme (which might include early corrective interventions), protective effects of both hips and spines might be included. This way of implementing early interventions may differ from other countries without follow-up programmes. The chosen definition of limited knee extension with a cut-off value of -5° or more affected the observed frequency of knee contracture. A higher cut-off value, for example -10° , would have halved the prevalence from 14% to 7%. A changed cut-off value for hip extension, which was removed due to non-significance in the multivariable analyses, could perhaps have changed the end result. We dichotomized epilepsy; another classification, for example, that of Bertocelli et al., might have altered the result. Due to the low number of Cobb angles reported in the registry, we chose to include both radiographic and clinical definitions of scoliosis. However, for clinical examination, there is high sensitivity and specificity compared with the radiographic Cobb angle. We had a low frequency of children with MP $>40\%$, which may have affected the result. MP could potentially be a risk factor in a population without hip surveillance. Finally, there might be other predictors that were not identified or included in this study.

Clinical implications: can science improve Kid's everyday life?



And here is Kid again, on his first try-out of a new soft spinal orthosis (but not yet in his new, customized wheel-chair). Behind the blanked-out face is a big smile, with a child saying “I never want to take the brace off”, and telling his Dad “Look, I can use my hands”, while he waves them around in the air. Also, his head is upright, making social interactions easier, and talking to us for much longer periods of time than he used to.

But in relation to the current thesis, is the new knowledge useful for Kid?

We know now that the risk for Kid to develop a scoliosis, even as an adult, is high, especially as he is classified at GMFCS level V. His risk for scoliosis will increase with age, while the fact that he is male decreases it slightly. As he will be monitored closely in the national follow-up programme for children with CP (CPUP), together with the possibility to continue with surveillance into adulthood, the risk to develop a severe scoliosis over time will likely decrease. Kid's use of a spinal orthosis also decreases the risk to develop scoliosis, as higher goal attainment levels are achieved for the goal of prevention of deformity when it is used as a preventive treatment before the development of moderate or severe scoliosis occurs. But Kid is one of those children who does not yet have a scoliosis but is provided a spinal orthosis primarily for functional reasons. There are

functional benefits that were apparent to both him and us already the first time he put on a brace. For Kid, all four CPUP goals are relevant: preventing deformity, and improving stability/positioning, arm–hand function, and head control. The proportion of children wearing a spinal orthosis is low, while the functional goals and goal attainment are high. Kid is now one of the children who benefits from improved stability, head control and arm–hand function. He also has the additional postural support provided by the spinal orthosis for long hours, which may prevent the development of scoliosis.

Can we predict Kid’s future risk of severe scoliosis before the age of 16? By using the risk score based on male sex, GMFCS V, not having epilepsy but having limited knee extension, the calculation shows that Kid’s individual risk to develop scoliosis is a score of 0.09, which translated into a risk score level increases the risk by 50–60% (exact value 52%). This knowledge may well encourage Kid’s current physiotherapist to offer interventions for his knee contractures, with the aim of reducing the risk for scoliosis further, together with the close surveillance of hips and spine that CPUP offers.

Because the CPCHILD was found to be a sound proxy-reported questionnaire for children with low motor functioning, it can now be used to evaluate the interventions given. For example, Kid’s current physiotherapist or orthopaedic surgeon can evaluate his HRQoL in relation to the intervention of a new spinal orthosis, a new wheel-chair or spinal surgery.

So the important question is: have these interventions changed Kid’s HRQoL?

As this story took place back in 2012 before I started my research, I already know the answer. Kid wore his brace most hours during the day, both at school and at home, and every morning he told his Dad that he wanted to put it on. (As Kid had no scoliosis, there were no specific hours during which he had to wear the brace.) After a couple of weeks, his new wheel-chair arrived and he immediately started moving himself around, both inside and outside, with an increased use of his arms. He liked the outdoor breaks at school, driving himself around the schoolyard, chasing friends in his wheel-chair. Before the brace and the new wheel-chair, he just sat where he had been placed. After Kid started using the brace, his teacher said that he interacted more and for longer periods of time with his classmates, teachers and staff, primarily because he could now hold his head up for long periods of time. As a clinician, I would have ticked all the boxes for “yes” for each goal and goal attainment, when evaluating the use of the brace. In this particular case, I believe that we achieved the ultimate treatment goal of improving Kid’s activity, participation and thereby QoL.

Att veta när man vet något och att veta när man inte vet något – det är kunskap

Konfucius 555 f.Kr.–479 f.Kr.

Conclusions

This thesis has described the development of moderate and severe scoliosis in individuals with CP; we identified predictors for severe scoliosis that form the basis of a risk score and explored the use of spinal orthoses in a total population of individuals with CP. We also evaluated an instrument to examine HRQoL that was developed for children with lower gross motor function, GMFCS levels IV and V.

The Scandinavian version of the CPCHILD for children with CP showed construct validity, high test–retest reliability and high internal consistency. Known-group validity for the total and domain scores of the CPCHILD differed significantly according to the children’s GMFCS level. Our findings confirm the initial hypothesis that children with lower motor ability (i.e., higher GMFCS level) would have lower scores on the CPCHILD. To provide effective care for children with disabilities, it needs to be comprehensive, well co-ordinated and continuous because such children require different care to those with an acute or temporary illness. The CPCHILD can be used in both clinical and research settings in the Scandinavian countries for children with CP. The CPCHILD thus appears to be a valid and reliable proxy-reported measure for HRQoL.

For scoliosis, a higher GMFCS level was a significant risk factor for the development of scoliosis; it also occurred at younger ages in individuals classified at a higher GMFCS level and the incidence of scoliosis continued to increase up to the age of 20–25 years. Females had a higher risk of moderate or severe scoliosis compared with males, with a HR of 1.4. At 20 years of age, ~75% of those at GMFCS level V had a Cobb angle $\geq 40^\circ$, highlighting the strong correlation between the individual’s GMFCS level and the development of scoliosis. This knowledge contributes to identifying the critical ages for surveillance and can serve as a basis for guidelines for treatment of scoliosis. Surveillance programmes should be based on age and GMFCS level, and should be initiated at a young age and continued into adulthood.

Although the goal of using spinal orthoses to prevent curvature progression remains important, we found that their functional benefits (improved stability, head control and arm–hand function) were more common goals with higher goal attainment. Spinal orthoses were used by 9% of children with CP. They were equally common in boys and girls, and their use increased with age and GMFCS level. Spinal orthoses were mostly used to improve functional outcomes, and the overall rate of goal attainment was high. Most children who use a spinal orthosis

do so to improve function rather than prevent deformity. Goal attainment levels were high for stability, head control and arm–hand function, indicating functional benefits for children in their daily lives. There appear to be higher goal attainment levels for prevention of deformity when spinal orthoses are used as a preventive treatment before the scoliosis has become moderate or severe. Because the ultimate treatment goal is to improve activity, participation and QoL, children with postural deficits should be given the opportunity to explore the functional benefits of a spinal orthosis. To our knowledge, this is the first study of a total population of children with CP using spinal orthoses.

A risk score was developed based on the following risk factors assessed at the age of 5 years: female sex, GMFCS levels IV and V, epilepsy and having limited knee extension, which were all identified as independent predictors for the development of severe scoliosis before the age of 16. The AUC of the resulting risk score was high, indicating high discriminatory ability for differentiating between individuals at high and low risk for development of scoliosis before the age of 16. The risk score may be useful when considering interventions to prevent or predict severe scoliosis in young children with CP and will hopefully provide clinicians with early insight regarding a child’s risk of developing scoliosis. This will allow them to differentiate high- from low-risk individuals, optimize the surveillance of each individual and, when indicated, allow timely interventions for those at risk for scoliosis. To our knowledge, this is the first study creating a risk score for development of severe scoliosis based on predictors identified in 5-year-old children with CP.

In short

Surveillance programmes for scoliosis in CP should be based on age and GMFCS level, and should be initiated at a young age and continued into adulthood. Information on the individual risk score can help to initiate and implement preventive interventions and strategies at an early stage, and thereby reduce the risk for scoliosis. Children with postural deficits at risk for scoliosis may benefit from treatment with spinal orthoses to improve function and in some cases prevent deformity. The CPCHILD questionnaire appears to be a valid and reliable proxy-reported measure for HRQoL.

Future research

- To validate the self-reported version of the CPCHILD questionnaire, so that the children's perception of their own HRQoL can be investigated.
- To apply the CPCHILD in the clinical setting and investigate HRQoL for a variety of interventions, and compare the results between individuals, other groups or other countries.
- To continuously investigate the incidence and prevalence of scoliosis as the CPUP population ages to increase our knowledge about scoliosis in adults with CP.
- To investigate further the functional aspects of a non-invasive scoliosis treatment like the use of spinal orthoses, also from a qualitative angle.
- It would be desirable to verify the external validity of the risk score for severe scoliosis in other CP populations.

Sammanfattning, summary in Swedish

Cerebral pares (CP) är den vanligaste orsaken till motorisk funktionsnedsättning hos barn och ungdomar och årligen föds ca 200 barn med CP i Sverige. Symtomen vid CP varierar, men karaktäriseras av bland annat nedsatt rörelseförmåga och nedsatt förmåga att stabilisera kroppen. Nästan åtta av tio vuxna med CP besväras av korta och spända muskler, snedställda leder och smärta som påverkar deras funktion och livskvalitet. Utan strukturerad uppföljning och förebyggande tidiga insatser, utvecklar vart fjärde barn med CP skolios som kan ha stor inverkan på deras livskvalitet. Orsaken till skolios är inte helt klarlagd och det är därför av stor vikt att identifiera riskfaktorer för att kunna förutsäga vilka barn med CP som löper risk att utveckla en behandlingskrävande skolios. Vissa studier tyder på att korsettbehandling till viss del kan bromsa upp skoliosens försämringshastighet och underlätta funktion, men kunskapsläget är oklart. Personer med CP rapporterar ibland lägre livskvalitet. För att kunna utvärdera hälsorelaterad livskvalitet hos de barn som har svårast form av CP, har ett frågeformulär (CPCHILD) utvecklats av barnortopedier i Kanada.

The Gross Motor Classification System (GMFCS) är ett standardiserat klassifikationssystem som delar in och beskriver motorisk förmåga i fem nivåer hos individer med CP, där individer i nivå I har högst och individer i nivå V har lägst motorisk förmåga. Studie II - IV, använde sig av data från CPUP (Cerebral Pares Uppföljnings Program), ett svenskt uppföljningsprogram för personer med CP som startade 1994 och som sedan 2005 även är ett nationellt kvalitetsregister.

Syftet med avhandlingen var att undersöka skoliosutveckling och korsettbehandling hos personer med CP, att identifiera riskfaktorer för att utveckla skolios samt att utvärdera frågeformuläret CPCHILD för svenska och norska förhållanden.

Studie I - Familjer i Sverige och Norge bjöds in för att delta i utvärderingen av CPCHILD (Caregiver Priorities and Child Health Index of Life with Disabilities) och 123 familjer tackade ja till att delta. Validitet (att enkäten mäter det den avser att mäta) och test-retest reliabilitet, (att enkäten mäter samma sak vid upprepad mätning) utvärderades. CPCHILD uppvisade hög validitet samt förmåga att särskilja mellan GMFCS nivåer och en hög tillförlitlighet avseende test-retest reliabilitet.

Studie II – Undersökning av förekomsten av måttlig och uttalad skolios i förhållande till kön, GMFCS nivå och ålder, hos 1025 individer med CP från södra Sverige. Förekomsten av skolios ökade med ålder och med ökad GMFCS nivå (sämre motorisk förmåga). Flickor hade en något ökad risk för skolios. Skoliosutvecklingen började tidigare hos individer med hög GMFCS nivå och skoliosen fortsatte utvecklas upp till 20-25 års ålder.

Studie III – Undersökning av korsett användning hos barn med CP i Sverige (födda 2000-2014), samt analys av mål och målluppfyllelse för korsettbehandling i förhållande till ålder, kön, GMFCS nivå och förekomst av skolios. Nio procent av 2800 barn använde korsett, där användandet ökade med stigande ålder. Bara barn i GMFCS nivå III-V använde korsett och användandet ökade med högre GMFCS-nivåer. Av de som korsettbehandlades hade 59% skolios. De flesta barn rapporterade flera mål, där förbättrad funktion var det vanligaste behandlingsmålet (att förbättra stabilitet, arm-handfunktion och huvudkontroll). En hög andel, 78-87%, ansåg att målen uppfylldes. Endast en tredjedel använde korsett för att förebygga skolios. Detta är den första studie som analyserat korsett användning hos en hel population barn med CP.

Studie IV – Syftet var att skapa en risk score för att förutse uttalad skolios före 16-års ålder. Vi fann 4 faktorer som vid 5 års ålder medförde en ökad risk för skolios; GMFCS nivå IV och V, att vara flicka, epilepsi samt nedsatt förmåga att sträcka i knäleden. Risk scoren uppvisar hög förmåga att särskilja mellan individer med hög kontra låg risk för att utveckla uttalad skolios. Detta är den första studien som utvecklat en risk score för uttalad skolios baserad på faktorer hos 5-åriga barn med CP.

SLUTSATSER:

Uppföljningsprogram för skolios vid CP bör baseras på ålder och GMFCS-nivå. Programmet bör påbörjas i ung ålder och fortsätta upp i vuxen ålder.

Den individuella riskscoren för skolios bör kunna bidra till att initiera och genomföra förebyggande interventioner i ett tidigt skede av skoliosutvecklingen.

Barn med CP med nedsatt förmåga att stabilisera kroppen samt barn med skolios, framförallt barn med lätt skolios, bör ges möjlighet att prova korsettbehandling.

CHILD är ett föräldraskattat undersökningsformulär som har en hög tillförlitlighet för att mäta häso relaterad livskvalitet hos barn med CP.

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Appendix

SEKTION 1: PERSONLIG VÅRD/AKTIVITETER I DAGLIGA LIVET

BEHOV AV ASSISTANS

								T O T A L	M Å T T L I G	Ö V E R V A K A D / M Å T T L I G	S J Å L V S T Å N D I G
<p>Tänk igenom hur var och en av följande aktiviteter vanligtvis utförs av/för ditt barn.</p> <p>Bedöm hur svårt var och en av dessa aktiviteter var under de senaste två veckorna, och ange hur mycket assistans (hjälp) ditt barn behövde för att utföra dessa aktiviteter.</p>											
Under de senaste 2 veckorna, hur svårt var följande:	<i>Inte möjligt</i>	<i>Mycket svårt</i>	<i>Svårt</i>	<i>Viss svårighet</i>	<i>Lätt</i>	<i>Mycket lätt</i>	<i>Inget problem</i>				
1. äta/dricka eller matas? (på det sätt det vanligtvis sker, t ex. via mun, peg eller båda)	0	1	2	3	4	5	6	0	1	2	3
2. upprätthålla munhygien? (hålla mun och tänder rena)	0	1	2	3	4	5	6	0	1	2	3
3. badning/tvättning?	0	1	2	3	4	5	6	0	1	2	3
4. toalettbesök? (blås och tarmfunktion, hygien etc)	0	1	2	3	4	5	6	0	1	2	3
5. byta blöjor/underkläder?	0	1	2	3	4	5	6	0	1	2	3
6. sätta på/ta av kläder på överkroppen? (skjorta, jacka etc)	0	1	2	3	4	5	6	0	1	2	3
7. sätta på/ta av kläder på underkroppen? (byxor etc)	0	1	2	3	4	5	6	0	1	2	3
8. sätta på/ha något på fötterna? (strumpor, skor, ortoser etc)	0	1	2	3	4	5	6	0	1	2	3
9. hårvård? (tvätta, torka, borsta/kamma, fläta etc.)	0	1	2	3	4	5	6	0	1	2	3
1A. annan aktivitet inom personlig vård? Specificera: _____	0	1	2	3	4	5	6	0	1	2	3
1B. annan aktivitet inom personlig vård? Specificera: _____	0	1	2	3	4	5	6	0	1	2	3

SEKTION 2: POSITIONERING, ÖVERFLYTTNING & FÖRFLYTTNING

BEHOV AV ASSISTANS

<p>Tänk igenom hur var och en av följande aktiviteter vanligtvis utförs av/för ditt barn.</p> <p>Bedöm hur svårt var och en av dessa aktiviteter var under de senaste två veckorna, och ange hur mycket assistans (hjälp) ditt barn behövde för att utföra dessa aktiviteter.</p>								T O T A L	M Å T T L I G	Ö V E R V A K A D /	S J Å L V S T Å N D I G																																																																																				
<p>Under de senaste 2 veckorna, hur svårt var följande:</p> <table style="width: 100%; border-collapse: collapse;"> <thead> <tr> <th style="width: 10%;"></th> <th style="width: 10%; text-align: center;"><i>Inte möjligt</i></th> <th style="width: 10%; text-align: center;"><i>Mycket svårt</i></th> <th style="width: 10%; text-align: center;"><i>Svårt</i></th> <th style="width: 10%; text-align: center;"><i>Viss svårighet</i></th> <th style="width: 10%; text-align: center;"><i>Lätt</i></th> <th style="width: 10%; text-align: center;"><i>Mycket lätt</i></th> <th style="width: 10%; text-align: center;"><i>Inget problem</i></th> </tr> </thead> <tbody> <tr> <td style="padding: 5px;">10. ta sig i och ur sängen? _____</td> <td style="text-align: center;">0</td> <td style="text-align: center;">1</td> <td style="text-align: center;">2</td> <td style="text-align: center;">3</td> <td style="text-align: center;">4</td> <td style="text-align: center;">5</td> <td style="text-align: center;">6</td> </tr> <tr> <td style="padding: 5px;">11. ta sig i och ur rullstol/stol? _____</td> <td style="text-align: center;">0</td> <td style="text-align: center;">1</td> <td style="text-align: center;">2</td> <td style="text-align: center;">3</td> <td style="text-align: center;">4</td> <td style="text-align: center;">5</td> <td style="text-align: center;">6</td> </tr> <tr> <td style="padding: 5px;">12. sitta i rullstol/stol? _____</td> <td style="text-align: center;">0</td> <td style="text-align: center;">1</td> <td style="text-align: center;">2</td> <td style="text-align: center;">3</td> <td style="text-align: center;">4</td> <td style="text-align: center;">5</td> <td style="text-align: center;">6</td> </tr> <tr> <td style="padding: 5px;">13. stå vid träning/överflyttning? _____</td> <td style="text-align: center;">0</td> <td style="text-align: center;">1</td> <td style="text-align: center;">2</td> <td style="text-align: center;">3</td> <td style="text-align: center;">4</td> <td style="text-align: center;">5</td> <td style="text-align: center;">6</td> </tr> <tr> <td style="padding: 5px;">14. förflytta sig i hemmet? (på vilket sätt som helst) _____</td> <td style="text-align: center;">0</td> <td style="text-align: center;">1</td> <td style="text-align: center;">2</td> <td style="text-align: center;">3</td> <td style="text-align: center;">4</td> <td style="text-align: center;">5</td> <td style="text-align: center;">6</td> </tr> <tr> <td style="padding: 5px;">15. förflytta sig utomhus? (på vilket sätt som helst) _____</td> <td style="text-align: center;">0</td> <td style="text-align: center;">1</td> <td style="text-align: center;">2</td> <td style="text-align: center;">3</td> <td style="text-align: center;">4</td> <td style="text-align: center;">5</td> <td style="text-align: center;">6</td> </tr> <tr> <td style="padding: 5px;">16. ta sig i och ur ett motorfordon? (bil, van, buss) _____</td> <td style="text-align: center;">0</td> <td style="text-align: center;">1</td> <td style="text-align: center;">2</td> <td style="text-align: center;">3</td> <td style="text-align: center;">4</td> <td style="text-align: center;">5</td> <td style="text-align: center;">6</td> </tr> <tr> <td style="padding: 5px;">17. besöka offentliga platser? (park, teater, sightseeing, etc) _____</td> <td style="text-align: center;">0</td> <td style="text-align: center;">1</td> <td style="text-align: center;">2</td> <td style="text-align: center;">3</td> <td style="text-align: center;">4</td> <td style="text-align: center;">5</td> <td style="text-align: center;">6</td> </tr> <tr> <td style="padding: 5px;">2A. annan aktivitet? Specificera: _____ _____</td> <td style="text-align: center;">0</td> <td style="text-align: center;">1</td> <td style="text-align: center;">2</td> <td style="text-align: center;">3</td> <td style="text-align: center;">4</td> <td style="text-align: center;">5</td> <td style="text-align: center;">6</td> </tr> <tr> <td style="padding: 5px;">2B. annan aktivitet? Specificera: _____ _____</td> <td style="text-align: center;">0</td> <td style="text-align: center;">1</td> <td style="text-align: center;">2</td> <td style="text-align: center;">3</td> <td style="text-align: center;">4</td> <td style="text-align: center;">5</td> <td style="text-align: center;">6</td> </tr> </tbody> </table>													<i>Inte möjligt</i>	<i>Mycket svårt</i>	<i>Svårt</i>	<i>Viss svårighet</i>	<i>Lätt</i>	<i>Mycket lätt</i>	<i>Inget problem</i>	10. ta sig i och ur sängen? _____	0	1	2	3	4	5	6	11. ta sig i och ur rullstol/stol? _____	0	1	2	3	4	5	6	12. sitta i rullstol/stol? _____	0	1	2	3	4	5	6	13. stå vid träning/överflyttning? _____	0	1	2	3	4	5	6	14. förflytta sig i hemmet? (på vilket sätt som helst) _____	0	1	2	3	4	5	6	15. förflytta sig utomhus? (på vilket sätt som helst) _____	0	1	2	3	4	5	6	16. ta sig i och ur ett motorfordon? (bil, van, buss) _____	0	1	2	3	4	5	6	17. besöka offentliga platser? (park, teater, sightseeing, etc) _____	0	1	2	3	4	5	6	2A. annan aktivitet? Specificera: _____ _____	0	1	2	3	4	5	6	2B. annan aktivitet? Specificera: _____ _____	0	1	2
	<i>Inte möjligt</i>	<i>Mycket svårt</i>	<i>Svårt</i>	<i>Viss svårighet</i>	<i>Lätt</i>	<i>Mycket lätt</i>	<i>Inget problem</i>																																																																																								
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15. förflytta sig utomhus? (på vilket sätt som helst) _____	0	1	2	3	4	5	6																																																																																								
16. ta sig i och ur ett motorfordon? (bil, van, buss) _____	0	1	2	3	4	5	6																																																																																								
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2B. annan aktivitet? Specificera: _____ _____	0	1	2	3	4	5	6																																																																																								

SEKTION 3: KOMFORT & KÄNSLOR

Under de senaste 2 veckorna , hur ofta upplevde ditt barn smärta eller obehag?							INTENSITET			
	<i>Varje dag</i>	<i>Väldigt ofta</i>	<i>Ganska ofta</i>	<i>Ett fåtal gång</i>	<i>En eller två gånger</i>	<i>Ingen gång</i>	S V Å R	M Å T L I G	M I L D	I N G E N
18. medan det äter/dricker eller matas? _____	0	1	2	3	4	5	0	1	2	3
19. vid toalettbesök? (blås & tarmfunktion, hygien, blöjbyte etc) _____	0	1	2	3	4	5	0	1	2	3
20. vid på-/avklädning? _____	0	1	2	3	4	5	0	1	2	3
21. vid förflyttningar eller lägesförändringar? _____	0	1	2	3	4	5	0	1	2	3
22. i sittande? _____	0	1	2	3	4	5	0	1	2	3
23. i sängliggande? _____	0	1	2	3	4	5	0	1	2	3
24. som stör ditt barns sömn? _____	0	1	2	3	4	5	0	1	2	3
3A. under annan aktivitet? Specifera: _____ _____	0	1	2	3	4	5	0	1	2	3
3B. under annan aktivitet? Specifera: _____ _____	0	1	2	3	4	5	0	1	2	3
Under de senaste 2 veckorna, hur ofta var ditt barn:										
25. irriterad, upprörd eller arg? _____	0	1	2	3	4	5	0	1	2	3
26. olycklig eller ledsen? _____	0	1	2	3	4	5	0	1	2	3

SEKTION 4: KOMMUNIKATION & SOCIAL INTERAKTION

Tänk igenom hur var och en av följande aktiviteter **vanligtvis** utförs av/för ditt barn.

Bedöm hur **svår** var och en av dessa aktiviteter var under de senaste två veckorna.

Under de senaste 2 veckorna, hur stora svårigheter hade ditt barn:	<i>Inte möjligt</i>	<i>Mycket svårt</i>	<i>Svårt</i>	<i>Viss svårighet</i>	<i>Lätt</i>	<i>Mycket lätt</i>	<i>Inget problem</i>
27. att förstå dig?	0	1	2	3	4	5	6
28. att bli förstådd av dig?	0	1	2	3	4	5	6
29. att kommunicera med dem som inte känner ditt barn väl?	0	1	2	3	4	5	6
30. att leka själv?	0	1	2	3	4	5	6
31. att leka med andra?	0	1	2	3	4	5	6
32. att vara i skola/förskola?	0	1	2	3	4	5	6
33. att delta fritidsaktiviteter (simning, samvaro med familj och vänner, etc.)?	0	1	2	3	4	5	6
4A. annan social aktivitet? Specificera: _____	0	1	2	3	4	5	6
4B. annan social aktivitet? Specificera: _____	0	1	2	3	4	5	6

SEKTION 5: HÄLSA

Under de senaste 2 veckorna	<i>Var vänlig och ringa in det alternativ som stämmer bäst</i>					
34. Hur många gånger har ditt barn varit tvunget att besöka läkare eller sjukhus?	<i>Inlagd >7 dagar</i>	<i>Inlagd < 7 dagar</i>	<i>3 eller fler gånger</i>	<i>2 gånger</i>	<i>1 gång</i>	<i>Ingen gång</i>
	0	1	2	3	4	5

Under de senaste 2 veckorna	<i>Mycket Dåligt</i>	<i>Dåligt</i>	<i>Skapligt</i>	<i>Bra</i>	<i>Mycket Bra</i>	<i>Utmärkt</i>
35. Hur skulle du skatta ditt barns allmänna hälsotillstånd?	0	1	2	3	4	5

36. Skriv upp de mediciner ditt barn har tagit de senaste 2 veckorna

0. Inga mediciner

1. _____
2. _____
3. _____
4. _____
5. _____
6. _____
7. _____
8. _____
9. _____

SEKTION 6: DITT BARNS LIVSKVALITET

Under de senaste två veckorna	<i>Mycket Dålig</i>	<i>Dålig</i>	<i>Skaplig</i>	<i>Bra</i>	<i>Mycket Bra</i>	<i>Utmärkt</i>
37. Hur skulle du skatta ditt barns generella livskvalitet?	0	1	2	3	4	5

SEKTION 7: FAKTORER AV BETYDELSE FÖR DITT BARNS LIVSKVALITET

Hur stor betydelse tror du ditt barns nuvarande status beträffande varje enskild punkt nedan har för hans/hennes livskvalitet?	<i>Minst betydelse</i>	<i>Ingen stor betydelse</i>	<i>Viss betydelse</i>	<i>Måttlig betydelse</i>	<i>Stor betydelse</i>	<i>Störst betydelse</i>
1. Äta / dricka eller matas	0	1	2	3	4	5
2. Upprätthålla munhygien	0	1	2	3	4	5
3. Badning / tvättning	0	1	2	3	4	5
4. Toalettbesök / hygien	0	1	2	3	4	5
5. Byta blöjor /underkläder	0	1	2	3	4	5
6. På- /avklädning överkropp	0	1	2	3	4	5
7. På/avklädning underkropp	0	1	2	3	4	5
8. Sätta på / ha något på fötterna	0	1	2	3	4	5
9. Hårvård /skötsel	0	1	2	3	4	5
10. Ta sig i och ur sängen	0	1	2	3	4	5
11. Ta sig i och ur rullstol / stol	0	1	2	3	4	5
12. Sitta i rullstol / stol	0	1	2	3	4	5
13. Stå vid träning / överflyttning	0	1	2	3	4	5
14. Förflytta sig inomhus	0	1	2	3	4	5
15. Förflytta sig utomhus	0	1	2	3	4	5
16. Ta sig i och ur ett motorfordon	0	1	2	3	4	5
17. Besöka offentliga platser	0	1	2	3	4	5
18. Komfort vid ätande	0	1	2	3	4	5
19. Komfort vid toalettbesök	0	1	2	3	4	5
20. Komfort vid på-/avklädning	0	1	2	3	4	5
21. Komfort vid förflyttningar eller lägesförändringar	0	1	2	3	4	5
22. Komfort i sittande	0	1	2	3	4	5
23. Komfort i liggande	0	1	2	3	4	5
24. Komfort vid sömn	0	1	2	3	4	5
25. Känslostillstånd eller beteende	0	1	2	3	4	5
26. Lycka	0	1	2	3	4	5
27. Kan förstå dig	0	1	2	3	4	5
28. Kan bli förstådd av dig	0	1	2	3	4	5
29. Kan kommunicera med andra	0	1	2	3	4	5
30. Kan leka själv	0	1	2	3	4	5
31. Kan leka med andra	0	1	2	3	4	5
32. Kan vara i skola/förskola	0	1	2	3	4	5
33. Kan delta i fritidsaktiviteter	0	1	2	3	4	5
34. Minimera läkarbesök och sjukhusvistelser	0	1	2	3	4	5
35. Allmän hälsa	0	1	2	3	4	5
36. Minska antalet mediciner	0	1	2	3	4	5

SEKTION 8: (FAKTA) INFORMATION OM DITT BARN

1. Mitt barn är en:	<input type="checkbox"/> Pojke <input type="checkbox"/> Flicka
2. Vilket är ditt barns födelsedatum?	____ / ____ / ____ Dag Månad År
3. Vilken är den <u>högsta</u> skolnivå ditt barn fullföljt? (Markera bara en nivå)	Förskola ____ Förskoleklass ____ Första klass ____ Andra klass ____ Tredje klass ____ Fjärde klass ____ Femte klass ____ Sjätte klass ____ Sjunde klass ____ Åttonde klass ____ Nionde klass ____ Gymnasium åk 1 ____ Gymnasium åk 2 ____ Gymnasium åk 3 ____

SEKTION 9: INFORMATION OM DIG

1. Är du:	<input type="checkbox"/> Man <input type="checkbox"/> Kvinna
2. Vilket är ditt födelsedatum?	____ / ____ / ____ Dag Månad År
3. Vilket av följande beskriver bäst din nuvarande jobbstatus (markera alla som passar)	Arbetar inte pga mitt barns hälsa ____ Arbetar inte av andra orsaker ____ Söker arbete utanför hemmet ____ Arbetar hel- eller deltid (antingen utanför hemmet eller i hemmabaserat företag) ____ Är hemmavarande på heltid ____
4. Vilket av följande beskriver bäst ditt förhållande till ditt barn?	Biologisk förälder ____ Styvförälder ____ Fosterförälder ____ Adoptivförälder ____ Vårdnadshavare ____ Professionell vårdgivare ____ Annat (förklara) _____
5. I genomsnitt hur många dagar per vecka är du ansvarig för omvårdnadsaktiviteter för ditt barn?	____ dagar per vecka
6. Vilken är den högsta utbildning du fullföljt?	Högstadium ____ Gymnasium ____ Högskola eller universitet ____ Annan utbildning ____

Hur lång tid tog det dig att fylla i detta frågeformulär? (minuter): _____

TACK FÖR DIN MEDVERKAN!