Hip and Spine in Cerebral Palsy

Persson-Bunke, Måns

2015

Hip and Spine in Cerebral Palsy

Måns Persson-Bunke

LUND
UNIVERSITY

DOCTORAL DISSERTATION
by due permission of the Faculty of Medicine, Lund University, Sweden.

To be defended at Lecturehall F2 in Blocket, Getingevägen 4, Skånes Universitetssjukhus, Lund. Friday, November 6th, 2015, 9.00 a.m.

Faculty opponent

Professor Hans Tropp
Linköping University, Dept of Orthopedics, Sweden.

Tutor
Professor Gunnar Hägglund

Co-tutors
Assoc. Professor Henrik Lauge-Pedersen and Ph.D. Elisabet Rodby-Bousquet
Lund University, Clinical Sciences, Dept of Orthopedics, Sweden

Organization

LUND UNIVERSITY

Dept. of Orthopedics, Clinical Sciences, Lund, Sweden

Author(s): Måns Persson-Bunke

DOCTORIAL DISSERTATION

Date of issue 2015-11-06

Title and subtitle : Hip and spine in cerebral palsy

Abstract*:*

Background: Children with cerebral palsy (CP) have an increased risk of scoliosis, contractures including windswept hip deformity (WS), and hip dislocation. In 1994, a follow-up program and registry for children and adolescents with CP (CPUP) was initiated in Sweden to allow the early detection and prevention of hip dislocations and other musculoskeletal deformities.

Purpose: To analyze the prevalence of scoliosis and WS in children with CP and to study the effect of CPUP. To evaluate the psychometric properties of screening methods to detect scoliosis and postural asymmetries in children with CP.

Methods: Studies I and II were cross-sectional studies of the total population of children and adolescents with CP in southern Sweden. Clinical and radiographical data from the CPUP registry were used to identify all children with WS and scoliosis. The impact of such hip surveillance and the preventive contracture program, CPUP, was analyzed. In studies III and IV, the interrater reliability and validity of the clinical spinal examination used in CPUP and of the Posture and Postural Ability Scale (PPAS) were evaluated in children and adolescents (6-16 years) with CP.

Results: The prevalence of WS decreased nonsignificantly from 12 to 7% but the numbers of children with WS, scoliosis and hip dislocation decreased significantly ($p<0.05$). It appears that the hip surveillance program has resulted in a reduction in the incidence of WS starting in the lower extremities but not in the incidence of WS starting with scoliosis. The prevalence of moderate or severe scoliosis was 11%. The risk of developing a moderate or severe scoliosis increased with Gross Motor Function Classification System (GMFCS) level and age. The clinical spinal assessment showed excellent interrater reliability (weighted kappa=0.96) and high concurrent validity compared with radiographic Cobb angle measurement. The sensitivity was 75%, and specificity was 99.8%. The sensitivity of the scoliometer measurement was 50% and the specificity was 91.7%. Clinical spinal assessment seems useful to screen for scoliosis in children with CP. The PPAS showed an excellent interrater reliability (kappa scores 0.77-0.99), high internal consistency, and construct validity. It can be used to detect postural asymmetries in children and adolescents with CP at all levels of gross motor function.

Conclusion: WS starting in the lower extremity and severe scoliosis seems to have been reduced by the hip surveillance program. The screening methods used for scoliosis and postural asymmetries appear valid and reliable.

Key words Cerebral palsy, windswept hip deformity, scoliosis, PPAS, psychometric evaluation, posture, children.

Classification system and/or index terms (if any)

Language ENGLISH

ISSN and key title 1652-8220

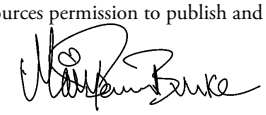
ISBN 978-91-7619-187-3

Recipient's notes

Number of pages

Price

Security classification

Signature  Date 2015-09-21

Hip and Spine in Cerebral Palsy

Måns Persson-Bunke

LUND
UNIVERSITY

Contact Address

Måns Persson-Bunke, MD

Department of Orthopedics

Skåne University Hospital

Lund, Sweden

Tel: +46 46 17 10 00

Mail: mans.persson-bunke@skane.se

Illustrations by Roland Van Veen

Lund University, Faculty of Medicine, Department of Clinical Sciences
Section of Orthopedics

Doctoral Dissertation Series 2015:108
ISBN 978-91-7619-187-3
ISSN 1652-8220

Printed in Sweden by Media-Tryck, Lund University
Lund 2015

When you can move less, a little is a lot

To all children and adolescents with cerebral palsy

Table of Contents

Original Papers

This thesis is based on the following papers, which will be referred to in the text by their Roman numerals.

I. Persson-Bunke M, Hägglund G, Lauge-Pedersen H. Windswept hip deformity in children with cerebral palsy. J Pediatr Orthop B 2006; 15:335-8.

II. Persson-Bunke M, Hägglund G, Lauge-Pedersen H, Wagner P, Westbom L. Scoliosis in a total population of children with cerebral palsy. Spine. 2012; 37: E 708-713.

III. Persson-Bunke M, Czuba T, Hägglund G, Rodby-Bousquet E. Psychometric evaluation of spinal assessment methods to screen for scoliosis in children and adolescents with cerebral palsy. Submitted.

IV. Rodby-Bousquet E, Persson-Bunke M, Czuba T. Psychometric evaluation of the Posture and Postural Ability Scale for children with cerebral palsy. Clin Rehabil. 2015 June 30. DOI: 10.1177/02692155 15593612.

Abbreviations

ADL	Activities of Daily Living
AUC	Area under the Curve
CI	Confidence Interval
CP	Cerebral Palsy
CPUP	Cerebral Palsy Follow-Up Program and National Quality Register
GMFCS	Gross Motor Function Classification System
HD	Hip Dislocation
LR	Likelihood Ratio
MP	Migration Percentage
PPAS	Posture and Postural Ability Scale
ROM	Passive Joint Range of Motion
SCPE	Surveillance of Cerebral Palsy in Europe
WS	Windswept Hip Deformity

Definitions

Cerebral Palsy	A group of permanent disorders of the development of movement and posture, causing activity limitation, which are attributed to nonprogressive disturbances occuring in the developing fetal or infant brain. The motor disorders are often accompanied by disturbances of sensation, perception, cognition, communication, and behavior, by epilepsy, and by secondary musculoskeletal problems (1).
Hip Dislocation	Reimers' migration percentage of 100% (2).
Posture	The configuration of the body. The position of the body segments in relation to each other, the supporting surface and the environment (3).
Postural Ability	The ability to stabilize the segments of the body in relation to each other, and to the supporting surface. The ability to control the center of gravity relative to the base of support during both static and dynamic conditions (3).
Scoliosis	A lateral deviation of the spine in the coronal plane.
Spasticity	A motor disorder characterized by a velocity dependent increase in tonic stretch reflex with exaggerated tendon jerks, resulting from hyperexcitability of the stretch reflex (4).
Windswept Hip Deformity	Describes an abduction and external rotation position of one hip with the opposite hip in adduction and internal rotation.

Abstract

Background

Cerebral palsy (CP) is an umbrella term that includes a heterogeneous group of childhood onset disorders of movement and posture that are life long. CP is a static encephalopathy that affects the immature brain. The overall prevalence of 2-3 per 1000 live births has remained unchanged over time (5). The anatomical site and severity of the lesion in the developing brain produce different clinical manifestations of motor impairments. Children with CP have an increased risk of scoliosis, contractures including windswept hip deformity (WS) and hip dislocation that are associated with age and with the severity of CP. Spasticity, weakness and muscle imbalance cause a lack of dynamic control in counteracting the deforming force of gravity. Inability to move increases the risk of sustained asymmetric postures that can increase the progression of deformities. In 1994 a follow-up program and registry for children and adolescents with CP (CPUP) was initiated in the south of Sweden, an area of approximately 1.3 million inhabitants. The main purpose of the CPUP was to prevent hip dislocations, severe contractures and deformities. The CPUP includes the total population of individuals with CP in the area born in 1990 or later. The hip surveillance component of CPUP includes a radiographic follow-up of the hips in children born in 1992 or later. Since 2011, adults with CP have been included in the CPUP.

Aims

I. To analyze the prevalence of scoliosis and WS in children with CP and to study the effect of the CPUP.

II. To evaluate the psychometric properties of clinical assessment tools to detect scoliosis and postural asymmetries in children with CP.

Methods

Studies ***I*** and ***II*** were cross-sectional studies of children and adolescents from a total population of those with CP. Clinical and radiographical data from the CPUP registry were used to identify all children with WS and scoliosis. The impact of the hip surveillance program was analyzed.

In studies ***III*** and ***IV***, three independent examiners evaluated the psychometric properties of clinical spinal examinations, scoliometer measurements and of the Posture and Postural Ability Scale (PPAS) in a selection of 28 and 29 children with CP.

Results

Paper I. The prevalence of WS decreased from 12 to 7% (nonsignificant) but the numbers of children with WS, scoliosis and hip dislocation decreased significantly ($p<0.05$). It seems that the hip surveillance program has resulted in a reduction in the incidence of WS starting in the lower extremity but not in the incidence of WS starting with scoliosis.

Paper II. The prevalence of moderate or severe scoliosis was 11%. The risk of developing a moderate or severe scoliosis increased with Gross Motor Function Classification System (GMFCS) level and age. The scoliosis was most commonly diagnosed after the age of 8 years. Children in GMFCS levels I or II had almost no risk of having a moderate or severe scoliosis by 18 years of age, whereas children in GMFCS levels IV or V had a 50% risk.

Paper III. The clinical spinal assessment used in the CPUP showed excellent interrater reliability (weighted kappa=0.96) and high concurrent validity compared to radiographic Cobb angle, with higher sensitivity (75% vs. 50%) and specificity (99.8% vs. 91.7%) than did scoliometer measurements. Clinical assessment appears to have been useful when screening for scoliosis in children with CP.

Paper IV. The PPAS showed an excellent interrater reliability (kappa scores 0.77-0.91), internal consistency (kappa scores 0.95-0.96) and construct validity ($p<0.01$) in separating known groups (GMFCS-levels II-V). It can be used for all levels of gross motor function to detect postural asymmetries in children and adolescents with CP.

Conclusions

The severity of WS and the frequency of scoliosis seem to have been reduced by the CPUP program. The screening methods for scoliosis and postural asymmetries are consistent and valid and seem appropriate to use among children with CP.

Sammanfattning- Summary in Swedish

Cerebral Pares (CP) är en övergripande benämning på ett rörelsehinder som orsakas av en skada som inträffat i en omogen hjärna. Graden av rörelsehinder varierar. Det föds cirka 200 barn årligen i Sverige med CP. Hög muskelspänning i kombination med muskelförsvagning och svårigheter att ändra ställning kan leda till att höften går ur led (höftledsluxation) och att ryggen blir sned (skolios). Ju större grad av rörelsehinder desto större risk är det att drabbas av höftledsluxation och skolios. Höftledsluxation inträffar ofta i tidig ålder och kan leda till skolios men höftledsluxation kan också vara sekundär till uttalad skolios. Detta kan leda till att en snedhet i bäckenet uppstår så att en ledstelhet och felställning (windswept) som kallas windsweptställda höfter kan utvecklas. Såväl höftledsluxation, skolios och windsweptställda höfter kan bland annat leda till smärtor, sitt- och ligg svårigheter samt ökad risk för trycksår. Sedan 1994 finns i Skåne och Blekinge ett uppföljningsprogram för alla barn med CP (CPUP). Sedan 2005 är CPUP ett nationellt kvalitetsregister där alla regioner och landsting deltar. Syftet med CPUP är att genom fortlöpande undersökningar av rörelseorganen kunna kan ge förebyggande behandling i rätt tid för att hindra att höften går ur led och att uttalade kontrakturer uppstår.

Syftet med denna avhandling var att studera hur vanligt windsweptställda höfter och skolios är hos barn och ungdomar med CP samt att utvärdera kliniska undersökningsmetoder som används för att upptäcka dessa tillstånd i tid.

Material: En population av 207 barn i ***studie I*** och 666 barn och ungdomar med CP i Skåne och Blekinge i ***studie II***. Ett urval av 28 respektive 29 barn och ungdomar med CP i Skåne i ***studie III*** och ***IV***.

Metod: Alla barn inom CPUP följs med standardiserade kliniska och röntgenologiska undersökningar av bland annat rygg och höft. Med hjälp av registerdata analyserades förekomsten av windsweptställda höfter samt skolios. I de två övriga studierna analyserades asymmetrier och förmågan att behålla och ändra ställning med ett kliniskt bedömningsinstrument, the Posture and Postural Ability Scale (PPAS), i liggande, sittande och stående. Förekomst av skolios vid framåtböjning och upprätt sittande klassificerades av tre erfarna undersökare oberoende av varandra. Vi analyserade hur väl undersökningarna mätte det vi avsåg att mäta (validitet) genom att jämföra den kliniska ryggundersökningen med ryggröntgen och mätning av storlek

på den eventuella kröken av ryggen och att utvärdera om PPAS kunde särskilja mellan grupper av barn med olika motorisk funktion (GMFCS-nivå).

Resultat: Förekomst av windswepställda höfter minskade från 12% till 7% (ej statistiskt signifikant) efter införandet av höftuppföljningsprogrammet. Om andelen barn som blev ”windswepta” på grund av förebyggande höftoperation, som utfördes för att hindra höften från att gå ur led exkluderades, var minskningen statistiskt signifikant. Förekomsten av skolios var 17% mild och 11% måttlig till uttalad skolios och var relaterad till ålder och grovmotorisk funktion. PPAS metoden visade stor noggrannhet och den kliniska ryggundersökningen hade utmärkt överenstämmelse mellan bedömmare och bra samstämmighet med röntgen.

Slutsatser: Förekomst av ”windswepta” höfter som börjar i nedre extremiteterna samt måttlig eller uttalad skolios verkar ha minskat hos de barn som ingår i höftpreventionsprogrammet inom CPUP. Metoderna för ryggundersökning och PPAS kan användas kliniskt för att identifiera skolios och asymmetrier i nacke, bål, bäcken, ben, och armar hos barn med CP.

Thesis at a Glance

	Questions	Methods	Results	Conclusions
I	What is the prevalence of windswept hip deformity (WS) in a total population of children with CP? What is the impact of the hip surveillance program (CPUP) on the prevalence of WS?	Cross sectional study of a total population of 207 children with CP aged 10 years. WS was defined as >50% difference in abduction or hip rotation between the left and right hips.	The frequency of WS was 12% in the control group and 7% in the study group following the hip prevention program. Children with WS in the study group had lower frequency of WS and hip dislocation.	Hip surveillance program reduced the incidence of WS starting in the lower extremities.
II	What is the prevalence of scoliosis in a total population of children with CP? What is the association with gross motor function, age and CP subtype?	Cross-sectional study of a total population of 666 children with CP aged 4-18 years.	17% had mild scoliosis and 11% had moderate or severe scoliosis. The risk for scoliosis increased with GMFCS-level and age.	Follow-up programs for the early detection of scoliosis should be based on the child's GMFCS level and age.
III	What are the psychometric properties of clinical spinal examinations and scoliometer measurements for children with CP?	The spine of 28 children (6-16 years) with CP was examined in sitting and in forward bending by three independent raters. The values were compared with the radiographic Cobb angle.	The clinical examination of the spine had an excellent interrater reliability and the validity compared with the Cobb angle was adequate.	The clinical examination method used in CPUP was adequate and the use of a scoliometer was not advantageous.
IV	What are the psychometric properties of the PPAS for children with CP?	Posture and postural ability was scored by three independent raters for 29 children with CP (6 -16 years). Construct validity was evaluated based on GMFCS levels II-V.	Excellent interrater reliability (kappa scores (0.77-0.99) and high internal consistency (0.95-0.96). The PPAS differed between GMFCS-levels ($p<0.01$).	The PPAS can detect postural asymmetries in children with CP at GMFCS levels II-V.

Introduction

Cerebral palsy

Cerebral palsy (CP) is the most common persistent motor disability in childhood. The prevalence is 2-3 cases per 1000 live births (5, 6). CP is a static encephalopathy that affects the immature brain. Even though the neurological injury is regarded as nonprogressive the secondary musculoskeletal manifestations of CP can be looked upon as progressive. The severity of impairments varies, and besides the musculoskeletal problems associated disabilities such as epilepsy, visual and cognitive impairment, speech and learning difficulties are associated with the severity of CP (5). CP is a lifelong condition, but the estimated life expectancy is only a little shorter than that of individuals without CP (5, 7). It is important that all possible interventions that are made to help individuals with CP also aim to improve the quality of life and participation in social activities as well as focusing on physical function.

Over time, different definitions and descriptions of CP have been used (8, 9). When the follow-up program and registry for children and adolescents with CP (CPUP) was initiated in Sweden in 1994 it was most commonly defined according to Mutch et al. (10), who defined CP as "an umbrella term covering a group of non-progressive, but often changing, motor impairments, syndromes secondary to lesions or anomalies of the brain arising in the early stages of development." This definition was used in papers I and II in this thesis. In the rest of this thesis we use the most recent and widely used definition, presented in 2006 by Rosenbaum et al. (1). Rosenbaum et al. described CP as "a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders are often accompanied by disturbances of sensation, perception, cognition, communication, and behavior, by epilepsy, and by secondary musculoskeletal problems." This definition covers the definition of CP by Mutch et al. (10), but also adds postural aspects. By using this wider definition of CP we do not risk missing any affected children. Since Rosenbaum et al.'s definition is the most commonly used we started to use it in the CPUP in 2007.

Classifications of CP subtype

Different classifications of CP into subtypes have been used throughout the years. This may be topographical (e.g., hemiplegia or diplegia) or based on neurological findings (e.g., spastic, ataxic). The Swedish classification by Hagberg et al. (11) has been used worldwide since the 1950s until recently. The Hagberg classification was used in ***Study*** I. This classification of CP subtypes is based on three dominating clinical signs. The three main types of neurological symptoms are spastic, dyskinetic and ataxic forms. The spastic type can be divided into hemiplegia, diplegia, or tetraplegia, determined by the degree of involvement of the limbs. Hemiplegia involves one side of the body. Diplegia is bilateral and affects the lower limbs more than the upper limbs. Tetraplegia is defined as a bilateral involvement of arms equal to or greater than in the legs. Ataxic CP is either a diplegic or simple congenital form. The dyskinetic form of CP can be divided into a dystonic or a mainly choreoathetotic type. If a dominating symptom is impossible to find, CP is referred to as a mixed form.

The Surveillance of Cerebral Palsy in Europe (SCPE) (12) has constructed a classification that is now used and has been adopted worldwide. According to the SCPE, CP can be classified into unilateral spastic, bilateral spastic, dyskinetic, ataxic and nonclassifiable CP. The dyskinetic form can be subdivided into dystonic and choreoathetotic CP. In this thesis, the SCPE classification was used in papers II-IV.

Classification of gross motor function

The classification of subtypes of CP is combined with the assessment of activities with special focus on gross motor function. Even when the symptoms of CP are described by subtypes, the subtype by itself is not a functional limitation. The main symptom of CP in children is the restriction of motor function. A useful development in the classification of CP is the introduction of the Gross Motor Function Classification System (GMFCS). GMFCS is a five-level ordinal scale used to describe and classify functional abilities in children with CP (13). GMFCS is based on the children's self-initiated movement focused on sitting, transport, and mobility. Level I describes the highest level of function and level V describes the lowest (Figure 1).

The interrater reliability and validity of the GMFCS is high (14). The individual GMFCS-level remains stable over time in most children with CP (15, 16). The expanded and revised version of the GMFCS (17) constitutes five age-bands up to 18 years of age. It was published in 2008 and has been used for the last two studies in this thesis. For a complete description of the GMFCS see Palisano et al. (13).

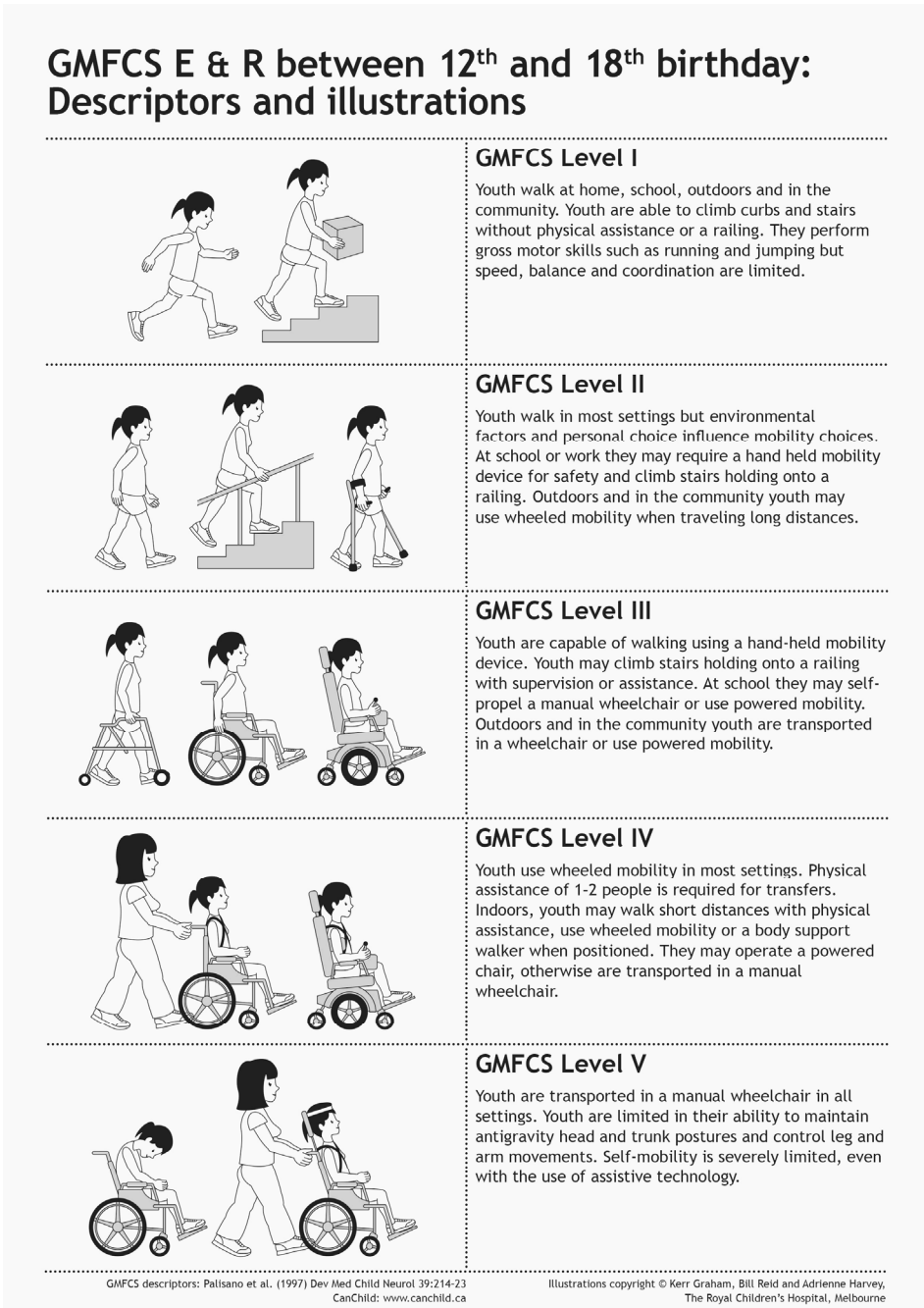


Figure 1.
Illustration of the GMFCS, with permission from Professor Kerr Graham.

CPUP: The follow up program for cerebral palsy

In 1994 a follow-up program and registry for children and adolescents with CP (CPUP) was initiated in the south of Sweden, an area with approximately 1.3 million inhabitants. CPUP has been classified as a National Healthcare Quality Registry in Sweden since 2005 and at present it is used in Norway, Denmark, Iceland, Scotland and New South Wales, Australia. In Sweden >95% of children with CP participate in CPUP (18).

CPUP is a secondary prevention program where the main priority is the prevention of hip dislocation, severe contractures, and deformities such as severe scoliosis and WS. It also aims to describe functioning and development in children wth CP as well as to evaluate treatment methods and to improve cooperation between health-care professionals. The National Healthcare Quality Registry gives opportunities for quality control, health -care planning and priority, and research (18).

The registry includes all children and adolescents born after January 1, 1990 living in the counties of Skåne and Blekinge, corresponding to a prevalence of 2.7 children with CP per 1000 (6, 18). Children born in 1992 or later are included in a hip surveillance program. All children with a suspicion of CP are included in the program as early as possible. The CP diagnosis and the neurological subtypes are confirmed by a neuropeditrician after 4 years of age. Children who do not fulfill the CP criteria at this age leave the program. The initial idea of the program was to prevent hip dislocation and contractures by a standardized follow up of children's hip and spine from an early age and to initiate preventive interventions if a deterioration was found.

Clinical examination

In the CPUP program, all children are examined regularly by their local physiotherapist and occupational therapist. The results are filled in to a recording form that includes measurements of passive ranges of motion (ROM) in all major joints with a goniometer. Children with GMFCS levels II-V are examined twice a year until the age of 6 and then once a year. Children with GMFCS level I are examined once a year until the age of 6 and then every second year.

The results from the clinical examinations are registered continuously and the team responsible for the treatment gets instant feedback so that preventive interventions can be initiated as soon as possible. Gross motor function is determined by the child's local physiotherapist according to their GMFCS level (13). A manual linked to the recording form describes the standardized measurements. Recommended values and intervals (critical values) from the measurements of passive ROM for different joints and for different GMFCS levels are graded like traffic light colors. Green means a good passive ROM. Yellow indicates a reduced ROM and need for increased

observation and to start actions to improve ROM. Red indicates the development of a contracture that requires intervention. It also is of importance to analyze the development of ROM over time.

The physiotherapist examines the spine with the child in a standing (in cases with compensation for a leg length discrepancy) or sitting upright position, with external support if needed, and then with forward bending. The degree of scoliosis is classified in the program as:

- No scoliosis.
- Mild: a discreet curve visible only on a thorough examination during forward bending.
- Moderate: an obvious curve visible during both extended and forward bending.
- Severe: a pronounced curve preventing the child from attaining an upright position without external support.

Radiographical examination

The hips are examined on anteroposterior radiographs with the subject in a supine position. The degree of displacement is measured as the migration percentage (MP) according to Reimers' (2) Figure 3. In children with GMFCS levels III-V the hips are examined from the time of enrollment and at least once a year until 8 years of age, and then on an individual basis. Children with GMFCS level II are examined at 2 and 6 years of age, and then on an individual basis. Children with GMFCS level I are not routinely examined radiographically unless a decreased ROM is discovered at clinical examination (www.CPUP.se). If fixed, moderate or severe scoliosis is found a radiographical examination is performed with the child sitting or standing. Further follow up depends on the Cobb angle and progression rate.

Treatments

The use of orthoses, orthopedic surgical treatment, serial casting or spasticity reducing treatment are also reported and documented.

The hip in cerebral palsy

In the hip, any muscle imbalance between strong and/or spastic hip flexors and adductors compared with weak extensor and abductor muscles, results in forces promoting lateral displacement of the femoral head (19, 20).

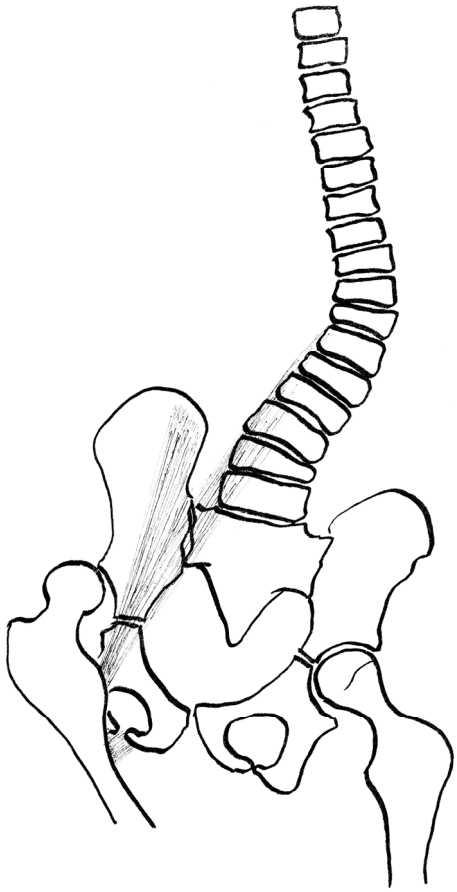


Figure 2.
Illustration of the possible origin of hip displacement in children with CP.

The development of hip dislocation usually starts at an early age and the rate of progression increases with age and with the severity of gross motor function impairment (21-23). Hip dislocation is often a painful condition that is difficult to treat. It is associated with scoliosis and contractures such as WS. It results in discomfort while sitting, standing, and lying down. Approximately 15% of all children with CP would sustain a hip dislocation unless preventive treatment is given (19, 21, 24, 25). The GMFCS level is useful as a guideline to predict those hips at risk for progressive lateral displacement (21). Soo et al. (24) found that the risk was 0% in children with GMFCS level I, and 90% in those with GMFCS level V.

In most cases, hip dislocation can be prevented by early detection and preventive treatment according to a hip surveillance program (22, 26-29).

A clinical examination is one component of the surveillance. This includes standardized and continuous ROM measurements with goniometer, clinical examination of the spine, and an assessment of spasticity.

Clinical examinations must be complemented with radiographical examination to identify hip displacement (21, 22, 30). For the radiographical follow-up, the measurement of Reimers' hip MP (2) is appropriate and probably the most used method (2, 21, 30). The MP describes the proportion of the femoral head that is positioned lateral to the acetabular margin (Figure 3).

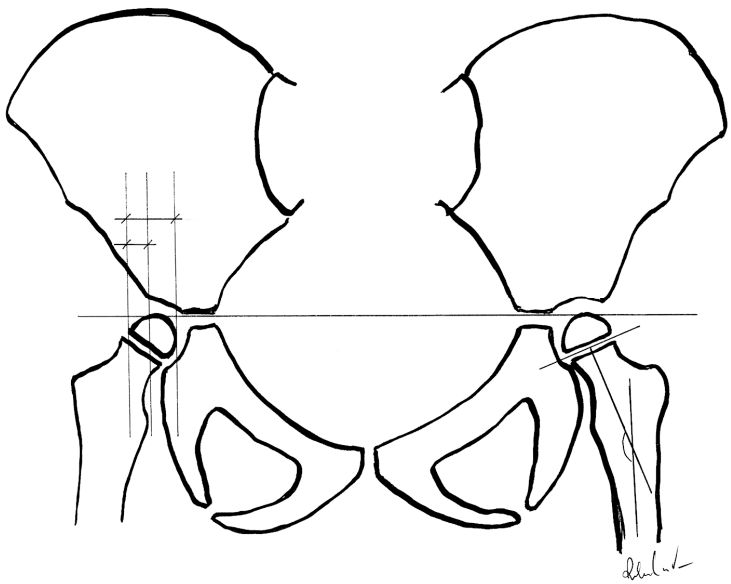


Figure 3.
Measurement of MP and femoral Head and Shaft angle.

Usually an MP >30-33% is defined as hip displacement and an MP of 100% as hip dislocation (2). In children with spastic CP the femoral head is often at a valgus angle in relation to the femoral neck. The head-shaft angle (Figure 3) also seems useful as a predictor for the risk of hip displacement in children with GMFCS levels III-V (31). This angle can be used as guidance and as a complement to age, GMFCS level, the MP and the clinical examination results when used for further interventions.

In the CPUP, standardized monitoring of the hips includes analyzing the development of MP over time. Children with an MP of <33% are not treated with preventive surgery, but proper positioning with the hips in abduction and extension is important. Children with MP values of 33-40 % are usually followed closely with a

special focus on risk factors for progression such as GMFCS level, age and head-shaft angle, and are treated surgically only when the MP increases.

Almost all hips with MP >40% will progress towards dislocation and the children often need surgical intervention (30). The choice of treatment method depends on the degree of dislocation, ROM, the degree of coxa valga and acetabular dysplasia, age and gross motor function. In the absence of a significant coxa valga and acetabular dysplasia, a bilateral adductor iliopsoas tenotomy might be sufficient if the MP is just above 40%. Unilateral soft tissue release is usually not recommended because it can increase the imbalance and cause a pelvic obliquity that will increase the risk of displacement of the opposite hip. One study has indicated that an adductor release might be more effective if a standing regime in abduction is performed postoperatively (32). If progression still occurs, a varization osteotomy of the proximal femur is required. In a study by Larsson et al. (33) a reoperation rate on the affected side of 25-30% was documented, highlighting the need for continuous follow–up and possibly the need for establishing the limits for acetabular dysplasia that motivates containment surgery on the pelvis. In cases of moderate or major acetabular dysplasia, a femoral varization osteotomy should be complemented with pelvic osteotomy to increase containment of the femoral head.

The spine in cerebral palsy

Scoliosis refers to a lateral deviation of the spine from the normal straight spinal alignment in the coronal plane (Figure 4).

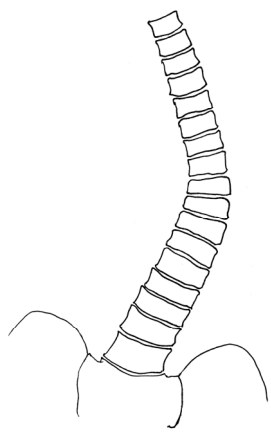


Figure 4.
Illustration of scoliosis.

Children and adolescents with CP have an increased risk of scoliosis and the reported prevalence is 15-64% according to study populations with different ages or severities of CP, and with different definitions of scoliosis (34, 35). In comparison the prevalence of adolescent idiopathic scoliosis is 2-4% (36, 37). In children with CP the risk of developing scoliosis is related to the child's gross motor function and age (35, 38).

Severe scoliosis is associated with pain, sitting problems, pelvic obliquity, hip dislocation, and WS. It can further impair cardiorespiratory function and quality of life, and might even be life threatening (35, 39-44). Scoliosis is the most common deformity of the spine, whereas deformities in the sagittal plane are not as common and are often regarded to be partially secondary to scoliosis (39).

The origin of scoliosis is not known completely, but can partly be caused by combinations of spasticity, asymmetric paraspinal muscle tone and strength, incoordination and imbalance, postural abnormality and the lack of dynamic control to oppose the forces of gravity (45).

Large scoliotic curves can be associated with other orthopedic problems such as hip dislocation, pelvic obliquity, and contractures such as WS (40), but the relationship is not fully understood (19, 46-48).

The speed of progression of scoliosis increases when the deforming forces of a large curve are reinforced by gravity. Scoliosis in children with CP can progress even after attaining skeletal maturity of the spine (38, 49). The larger the curve, the more likely it is for a scoliosis to progress and at a faster rate (38, 49). Saito et al. (38) concluded that the risk factors for progression were having a bilateral spastic involvement, being nonambulant and having a thoracolumbar curve. Curves of >40° before the age of 15 years progressed in 85% of the children studied.

Therefore, specific monitoring of scoliosis is analogous to hip surveillance, and it is important to detect it and identify any curve progression. The efficacy of spinal surgery is related to the curve's magnitude (42, 50).

A radiographic evaluation constitutes an anteroposterior view of the entire spine. Weight-bearing positions give a more true value and are more useful. Measurement of the Cobb angle (51) is the most commonly used method to measure the degree of scoliosis (Figure 5). On the radiographs the vertebrae with maximally tilted endplates below and above the apex are identified and the angle between the lines drawn along the superior and inferior endplates is defined as the Cobb angle.

There is an intraobserver error of 3-5° and an interobserver variability of 5-7° (52, 53) which has to be noted. There is no widely accepted grading classification of the degree of scoliosis (54).

The curves are named after the location of the apical vertebrae and are described as left or right depending on the shape of configuration. Whether the scoliosis is fixed or flexible is also often described. The curve pattern differs from idiopathic scoliosis and is often C- shaped with or without pelvic obliquity (55). (Figure 6).

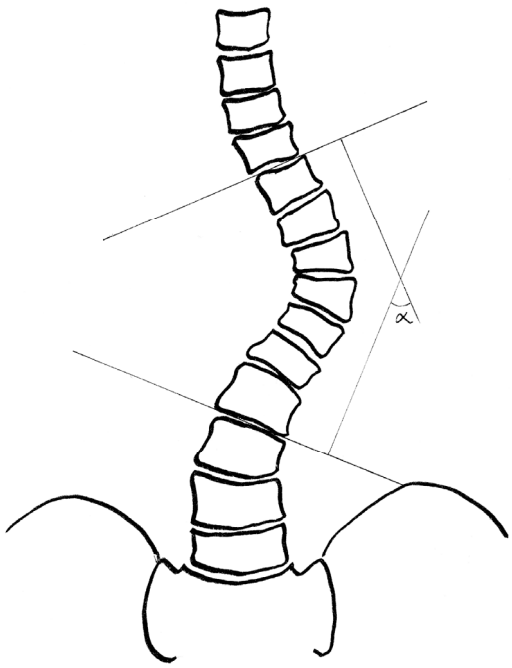


Figure 5.
Measurement of the Cobb angle.

Figure 6.
C-shaped scoliosis with pelvic obliquity and hip dislocation.

The goals of treatment are to reduce curve progression and improve function such as the subject's sitting ability, head control and balance, to reduce any pain from impingement of the ribs, and to prevent respiratory dysfunction.

Nonsurgical treatments include different sitting and postural supports, custom - molded seating inserts in wheelchairs and thoracolumbosacral orthoses (Figure 7) to support sitting, improve postural function, and try to reduce the rate of progression.

Figure 7.
Example of a custom molded spinal orthosis used to improve seating.

If the Cobb angle is used as an outcome measure, there is insufficient evidence for the effectiveness of brace treatment or a seating support to inhibit curve progression. The better the initial correction in the brace the slower is the rate of progression. The use of orthoses might have some effect on the Cobb angle if they are used for young children with flexible thoracolumbar or lumbar curves without excessive Cobb angles at the beginning of treatment (56-59).

Although the evidence for brace treatment on curve progression is insufficient some studies report functional benefits, subjective satisfaction, and ease of care. Spinal orthoses seem to improve seating, posture, balance, and associated control of the head, neck, and extremities in children with CP (59-61). The use of a brace does not seem to have any negative effect on pulmonary functions. A brace can even reduce the breathing workload according to Leopando et al. (62).

The indications for surgical treatment is progression of the scoliotic curvature that threatens to inhibit sitting or standing abilities (loss of function), or causes respiratory dysfunction, back pain, and pain because of pressure wounds from impingement of the ribs against the hemipelvis. A deformity with a Cobb angle >40-50° is usually an indication for surgical treatment (63). Outcome measures for operative treatment

include an improved Cobb angle, better respiratory function seating position and balance, enhanced activities of daily living (ADL), and reduced pain.

The possible benefits of surgical intervention have to be weighed against the risk factors and complications of surgery. Severely affected children can have poor nutrition, bad respiratory functions and an osteoporotic bone quality. There could also be reduced communication capacity and learning disabilities making it harder to receive information, and impair understanding and cooperation. The decision to perform surgery often lies with the carers and with no or limited participation from the patient. All individuals concerned in the decision-making have to consider the quality of life for the individual now with or without operative treatment currently and in the future.

Postoperative complications are more common than in surgery for idiopathic scoliosis (64). Early and late infections, pneumonia and respiratory failure, urinary tract infections, pseudarthrosis and implant failure are seen but the reported frequency varies between studies and from complications of methods not used any more (63, 65, 66).

The reported results of surgical treatment depend upon outcome measures in the different studies. If the Cobb angle before and after surgery is considered most studies show a positive result (67-71). In most studies, parents and caregivers report a high degree of satisfaction and functional benefits in ADL in spite of several complications (67-70, 72-75). In a retrospective study among 84 adolescents with CP followed on average 6 years postoperatively, Watanabe et al. (67) found an overall satisfaction rate of 92%. Better sitting balance was reported by 93% of the subjects and improvement in the quality of life by 71%.

Windswept hip deformity

Neither the spine nor the hip ought to be viewed in isolation. Sometimes called windblown syndrome, WS describes an adduction contracture and increased internal rotation of one hip and the other hip in abduction and external rotation (44, 76, 77). (Figure 8).

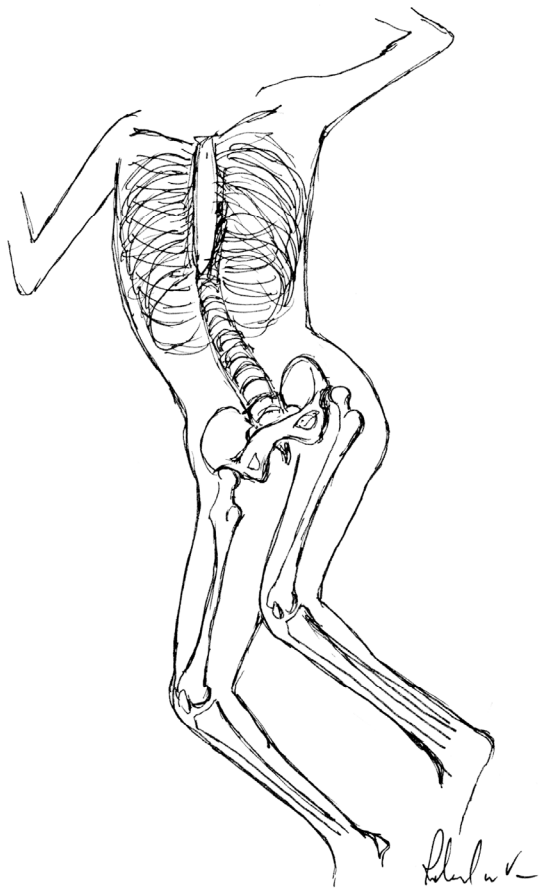


Figure 8.
Windswept hip deformity

WS is a clinical manifestation in some children with CP (44). The prevalence varies depending on how it is defined. In this thesis, WS is defined according to a modification of a formula constructed by Young et al. (77). Children with bilateral

CP and at least 50% difference in abduction, internal and/or external rotation between the left and right hip were defined as having WS. The risk increases with the severity of impairment and is a typical finding in children who are nonambulant (44, 77). WS is sometimes preceded by hip dislocation and sometimes by scoliosis, but the temporal relationship is unclear (44, 76). WS is a severe problem affecting weight distribution, pressure, and positioning in supine, prone, sitting, and standing positions. Pain and difficulties with hygiene and nursing care might arise. The aim of carers must be to prevent the development of WS. The hip and spine have to be monitored at an early age and preventive procedures for developing hip displacement and scoliosis must be undertaken. Correct positioning in lying, standing, and sitting might also help to prevent the development of WS (76). Hip or knee contractures can predispose to an asymmetric posture where the legs are swept to one side in lying and sitting positions and thereby induce a WS deformity (78).

Posture and postural ability

Posture is defined in this thesis as the configuration of the body. It covers the position of the body segments in relation to each other, the supporting surface and the environment (79). No uniform definition exists to our knowledge. The term posture is also included in the definition of CP by Rosenbaum et al. (1): "a group of permanent disorders of the development of movement and posture." A persistent deviation of the body from the midline might induce an asymmetric posture which in children with severe CP can result in contractures and bone and joint deformities, leading for example to scoliosis, pelvic obliquity, hip dislocation and windswept deformity (80-83).

Postural ability refers to the ability to stabilize the segments of the body in relation to each other and to the supporting ground during both static and dynamic situations. This means controlling the center of gravity relative to the base of support, and the ability to maintain and move into or out of different positions of the body (79, 84). Children with CP can have varying degrees of brain damage in the areas responsible for normal postural control and balance (85, 86). Damage to the brain stem, spinal cord, or basal ganglia can cause postural deficits. These vary from being unable to compensate against the force of gravity when the body deviates from midline equilibrium, or being unable to change position. Asymmetric postures may develop that can cause progressive deformities (80-82, 87). It is important to early identify postural asymmetries and deviations to prevent or minimize their consequences.

The Posture and Postural Ability Scale (PPAS)

The PPAS was designed to assess posture in people with severe disabilities, in terms of 'quality' of posture in terms of body shape and 'quantity' in terms of postural ability. It has excellent psychometric properties when evaluated for adults with CP (3). The PPAS consists of a seven-point ordinal scale for the assessment of postural ability in supine and prone lying, sitting, and standing; six items for assessing the quality of posture in the frontal plane; and six items in the sagittal plane (Table I). Good symmetry and alignment is scored 1 point for each item while asymmetry or deviation from the midline is scored 0 points. The total score for each position in the frontal and the sagittal plane is calculated separately. The two lower levels of ability are in fact rating no ability; i.e., inability to maintain or change position. The difference between these two levels is whether the person can (level 2) or cannot (level 1) conform to the position when placed by another person, i.e., in anatomical alignment with support. When a person cannot be placed in the specified position because of significant contractures and deformity, the postural ability is scored as level 1 meaning unplaceable and posture is scored 0 (89).

Table 1 The Posture and Postural Ability Scale (PPAS) with a seven-point ordinal scale for assessing postural ability in standing, sitting, supine and prone positions; followed by six items for assessing postural quality posture in the frontal plane; and another six items in the sagittal plane.

	PPAS Levels of postural ability
Level 1	Unplaceable in an aligned posture
Level 2	Placeable in an aligned posture but needs support
Level 3	Able to maintain position when placed but cannot move
Level 4	Able to initiate flexion/extension of trunk
Level 5	Able to transfer weight laterally and regain posture
Level 6	Able to move out of position
Level 7	Able to move into and out of position

PPAS Quality of posture , frontal view, (Yes = 1 point, No = 0 points)	
Standing, sitting, supine	**Prone**
Head midline	Head to one side
Trunk symmetrical	Trunk symmetrical
Pelvis neutral	Pelvis neutral
Legs separated and straight relative to pelvis	Legs separated and straight relative to pelvis
Arms resting by side	Arms resting (elevated, mid-position)
Weight evenly distributed	Weight evenly distributed

PPAS Quality of posture , sagittal view, (Yes = 1 point, No = 0 points)			
Standing	**Sitting**	**Supine**	**Prone**
Head midline	Head midline	Head midline	Trunk in neutral position
Trunk in neutral position	Trunk in neutral position	Trunk in neutral position	Pelvis neutral
Pelvis neutral	Pelvis neutral	Pelvis neutral	Hips extended
Legs straight, hips/knees extended	Hips mid-position (90°)	Legs straight, hips/knees extended	Knees extended
Feet mid-position/flat on floor	Knees mid-position (90°)	Feet resting in normal position	Arms resting (elevated, mid-position)
Weight evenly distributed	Feet mid-position/flat on floor	Weight evenly distributed	Weight evenly distributed

The purposes of this thesis

The aims of this thesis were to analyze the prevalence of scoliosis and WS in children with CP; to study the effect of the introduction of CPUP; and to evaluate clinical assessment tools used to screen for scoliosis and postural deficits in children with CP.

Study I: To study the prevalence of WS in a total population of children with CP, and to analyze what effect the introduction of the hip surveillance program and early treatments of contractures in the CPUP had on the prevalence of WS.

Study II: To describe the prevalence of scoliosis in a total population of children with CP, to analyze the relation between scoliosis, gross motor function, and CP subtype and to describe the age at diagnosis of scoliosis.

Study III: To evaluate the psychometric properties of the clinical spinal examination method used in CPUP.

Study IV: To evaluate the inter-reliability and construct validity of the PPAS in children and adolescents with CP.

Materials and Methods

Study designs

Studies **I** and **II** were cross-sectional studies of a total population of children with CP based on data collected from the CPUP registry. Study III evaluated the psychometric properties involved in the clinical examination method of the spine in CPUP. Study IV evaluated the PPAS in children and adolescents with CP.

Participants and methods

Study I included 207 children, with CP tracked by the CPUP and living in southern Sweden (Skåne och Bekinge). Only those who were born in the area or who moved into the area before 2 years of age and who were still living in the area at the age of 10 years were included.

Study II included 666 children, with CP in the regions of Skåne and Blekinge. Children participating in the CPUP and born between January 1, 1990, and December 31, 2004, were included. Children who died or who moved out of the area before the age of 5 years were excluded.

Study III included 28 children and adolescents with CP in Skåne (14 girls), aged 6 - 16 years. They were in GMFCS levels II (n=9), III (n=7), IV (n=6), and V (n=6).

Study IV included 29 children and adolescents with CP in Skåne (14 girls), aged 6 - 16 years. They were in GMFCS levels II (n=10), III (n=7), IV (n=6), and V (n=6).

In ***Study I,*** data were extracted from the CPUP registry. Of the 207 included participants, 68 were born in 1990-1991. They did not participate in the hip surveillance program and were regarded as a control group. The 139 children born in 1992-1995 were included in the hip surveillance program and constituted the study group.

Children with bilateral CP and at least 50% difference in abduction and internal or external rotation between the hips were defined as having WS. At least two

measurements in sequence with this difference were required. Those with a Cobb angle of 20° or more were regarded as having scoliosis. The frequency of WS, hip dislocation, scoliosis and those requiring proximal varization osteotomy were registered up to 10 years of age.

By using the same set of variables we analyzed this cohort again at the age of 20 years, although these are not published data.

In ***Study II,*** clinical and radiographical data from all children living in Skåne and Blekinge in the CPUP registry were used to identify children with scoliosis. This study was based on 7200 measurements in 666 children with CP aged 4 -18 years on January 1, 2008. The age at the first clinical diagnosis of scoliosis and the Cobb angle at the first radiographical examination were registered and analyzed in relation to GMFCS, age and CP subtype. The scoliosis was classified and graded according to the guidelines of CPUP (p. 23).

In ***Study III,*** 28 children aged 6 -16 years with CP participating in CPUP and in GMFCS levels II-V were recruited from five child rehabilitation units in southern Sweden. Children at GMFCS level I, which constitutes about 40% of all children with CP, do not have a higher risk for scoliosis than children with idiopathic scoliosis and were not included in this study (35). This reduced the number of children exposed to unnecessary radiation. The participants and their families were informed about the study by their local physiotherapist, and provided with invitation letters with information about the study. Written consent was collected from all participants. Children were recruited consecutively until at least six children at each GMFCS level had accepted. The reason for including six persons in each GMFCS level II-V was based on an earlier study evaluating the PPAS in adults with CP (3, 78). Three experienced raters examined each child once. The spine was examined clinically and with scoliometer measurement (88) with the children in a sitting position. The scoliometer was placed with the subject bending at the top of the thoracic spine, with the 0 (zero) mark over the spinous process, and slowly moved down the spine noting the highest degree of trunk rotation. Each rater noted the degree of scoliosis separately and independently. Higher grades indicate worse inclination and the value for defining scoliosis that needed radiographic examination was set to ≥7°. The results were compared with radiographic measurements of the Cobb angle and moderate or severe scoliosis was defined as a Cobb angle >20°. Radiographic examinations were performed with the children in a sitting position, on an anteroposterior projection.

In ***Study IV,*** 29 children and adolescents aged between 6 and 16 years were recruited at the same time according to the same procedure, principles and inclusion criteria as in Study III. The psychometric evaluation of the PPAS was completed at same occasion and by the same three raters as in Study III. All three raters had many years of experience working with children with CP but only one of them had experience of

rating posture and postural ability using the PPAS. The other two raters were given brief instructions before assessing the children. The children were instructed by one of the raters to get into and out of supine, prone, sitting positions on a plinth and into and out of a standing position on the floor. If they were unable to do this by themselves, they were placed in one of the positions and instructed to maintain it, initiate flexion of the trunk (when supine) or extension (when prone), transfer weight laterally and regain position, and move out of position, according to the levels of the PPAS (Table 1). If needed, the children were provided with manual support to stay in position. The children were also instructed to sit, stand or lie down in prone or supine positions as straight as possible, or were placed as straight as possible in the specified position and allowed to settle. The three raters assessed the posture and postural ability simultaneously and noted the scores on separate PPAS scoring sheets (Appendix).

Statistics

For the statistical analysis STATA (Stata Corp., College Station, TX, USA) and the R software environment (version 3.0.0; https://www.r-project.org) were used in studies **I** and **II**, and STATA (version 13.1) in studies **III** and **IV**; p -values less than 0.05 were considered significant for all statistical analyses.

In ***Study I*****,** Fisher's exact test (89) was used because of the small sample sizes, and because the data were categorical and binary in character.

In ***Study II***, linear regression estimates were used to evaluate the effect of age, CP subtype, and GMFCS levels on the magnitude of the Cobb angle at the first radiographical examination performed for diagnosing scoliosis. Data for subjects at GMFCS level I and with unilateral spastic CP were used as reference categories.

Kaplan-Meyer analysis was used to identify the age at diagnosis of moderate or severe scoliosis. The purpose was to illustrate the probability of NOT being diagnosed with scoliosis over time for subjects at different GMFCS levels.

Cox regression analysis was used to analyze the risk ratio (hazard ratio) for developing a clinical moderate or severe scoliosis in relation to the GMFCS level and CP subtype. Data for subjects at GMFCS level I and/or with spastic unilateral CP were used as reference categories.

In ***Study III,*** the interrater reliability for clinical spinal examination and scoliometer measurement were calculated using weighted kappa scores (90). The magnitude of weighted kappa was interpreted according to Fleiss 1981 where ≤0.40 signifies poor agreement, 0.40-0.75 fair to good agreement and ≥0.75 signifies excellent agreement

(91). To calculate the 95% confidence interval (CI) for weighted kappa scores, all GMFCS levels were combined and 95% nonparametric bootstrap CIs were added based on 1000 repeated samples (92, 93).

For evaluating concurrent validity, the Cobb angle was used as the gold standard. The area under a receiver operating characteristic curve (AUC), sensitivity, specificity, and predictive values were calculated. Averaged ratings were used for analyzing the validity of the scoliometer measures but not for calculation of kappa values.

The AUC measures the capacity of a test to classify a person correctly as being sick or not. In ***Study III***, the AUC was a measure of the capacity to identify a scoliosis correctly according to our definition. A value of <0.5 is not better than random, >0.7 is acceptable, >0.8 is excellent, and >0.9 is an extraordinary capability (89).

The likelihood ratio (LR) is a summary of the diagnostic accuracy of a test telling the ratio of the probability of a certain test result for individuals who do have the disease to the probability for individuals who do not. The definition of a positive LR is sensitivity/ 1 -specificity. The definition of a negative LR is 1-sensitivity/ specificity. A positive LR ≥10 means that the test is good at confirming scoliosis. A negative LR ≤0.2 means that the test is good at ruling out scoliosis (94).

In ***Study IV,*** interrater reliability was calculated using weighted kappa scores as in *Study III*. The magnitude of the weighted kappa scores indicates the agreement beyond chance. It was interpreted according to Fleiss 1981(91) as in *Study III*.

Construct validity was evaluated for known groups based on the GMFCS levels using the Jonckheere -Terpstra test for analyzing arithmetic average values given by the three raters.

Internal consistency was evaluated using Cronbach's alpha. This is a measure of item interrelatedness calculated with averaged values for the three raters, and Corrected Item-total correlation. It indicates the correlation between each item and the total score. Cronbach's alpha if item is deleted corresponds to the value achieved if a specific item is removed and the level should exceed 0.2 (95).

For evaluating of interrater reliability and internal consistency all GMFCS levels were combined and 95% nonparametric bootstrap CIs were generated based on a 1000 repeated samples (92, 93).

Ethics

Ethical approval was granted by the Medical Research Ethics Committee at Lund University for *studies **I** and **II*** (LU-433-99) and *studies **III** and **IV*** (467/2013).

Results

The results are described in detail in each published paper (see attachments).

Windswept hip deformity (***Study I***)

In the control group of 68 children (not included in the hip surveillance program) eight (12%) developed WS. Of these six also developed scoliosis and five developed hip dislocation before the age of 10 years (Table 2; cases 1-8). Hip dislocation was diagnosed before WS in four children. Scoliosis was diagnosed before or at the same time as WS in five children (Table 2).

In the study group of 139 children, 10 (7%) developed WS. Of these, four developed scoliosis but none developed hip dislocation (Table 2; cases 9-18). To prevent hip dislocation eight children in the study group were operated on with varization osteotomy of the proximal femur. In three of them this caused a decrease in the ROM in abduction, diagnosing them as having WS by the definition of this study. Scoliosis was detected before WS in three children.

The frequency of WS was related to lower levels of motor function. Eleven of 18 children were in GMFCS level V, six in GMFCS level IV, and one in GMFCS level III.

At a follow-up at 20 years of age (not included in the published *Study I*) a further four children in the control group and three in the study group developed WS (Table 3). Of the 25 children with WS at 20 years of age, nine developed WS at the same time or after scoliosis. In three children, WS developed after femoral varus osteotomy (Table 4). The number of children with WS starting in the lower extremities was significantly reduced in the study group ($p=0.028$) if the hips defined as showing WS after the varization osteotomy were excluded.

Table 2.
Characteristics of eight children in the control group (cases 1 -8), and of 10 children in the study group (cases 9 -18) with WS at the 10-year follow-up.

Case no	Year of birth	CP-subtype[1]	GMFCS	Age at WS	Age at scoliosis	Age at HD[2]	Age at op[3]
1	1990	D	III	9.5	-	-	-
2		D	IV	7.5	9.0	6.0	-
3		D	IV	8.5	5.0	-	-
4		D	IV	6.5	6.5	-	-
5		T	V	8.5	8.5	7.0	-
6		T	V	6.0	5.5	5.0	-
7		T	V	7.0	-	5.0	-
8		T	V	7.0	7.0	8.0	-
9	1992	D	IV	8.5	-	-	6.5
10		D	IV	7.5	3.0	-	7.0
11		Dy	IV	4.0	-	-	-
12		D	V	2.0	-	-	5.5
13		Dy	V	7.5	-	-	6.0
14		Dy	V	4.0	-	-	4.0
15		T	V	8.0	9.0	-	-
16		T	V	6.0	5.0	-	7.0
17		T	V	5.5	-	-	6.0
18		T	V	4.5	2.5	-	6.0

[1)] According to Hagberg et al. D, spastic diplegia; T, spastic tetraplegia; Dy, dystonic type. [2)] HD, hip dislocation. [3)] Op, Varization osteotomy of proximal femur.

A statistical comparison (Fisher's exact test) between control and study groups is shown in Table 3.

Table 3.
Numbers of children with WS and with WS in combination with scoliosis (S) and hip dislocation (HD) and femoral osteotomy (FO) in the control and study groups at 10 and 20 years of age.

	Control group*	%	Study group*	%	p
WS 10	8	12	10	7	0.20
WS 20	12	18	13	9	0.071
WS + S 10	6	9	4	3	0.067
WS + S 20	10	15	11	7	0.10
WS + HD 10	5	7	0	0	0.003
WS + HD 20	5	7	0	0	0.003
WS + FO 10	0	0	8	6	0.04
WS + FO 20	1	2	9	7	0.10
WS + S + HD 10	4	6	0	0	0.011
WS + S + HD 20	5	6	0	0	0.004
WS + S + FO 10	0	0	4	3	0.20
WS + S + FO 20	1	2	8	6	0.14

*All calculations are based on the number of children at the 10-year follow-up (Control group n= 68, Study group n = 139).

Table 4.
Numbers of children with WS associated with the deformity first identified. FO; femoral osteotomy, S; scoliosis, HD; hip dislocation.

	Control group	%	Study group	%
WS first	4	6	5	4
HD first	4	6	0	0
FO first	0	0	3	2
S first	4	4	5	4

Scoliosis (*Study II*)

In the total population of 666 children and adolescents with CP, 192 (28%) had scoliosis according to the physiotherapist's reports in the CPUP registry.

The scoliosis was graded as mild in 116 children (17%), and was not examined radiographically according to the follow-up program guidelines. In the remaining 76 children (11%), the types of scoliosis were graded as moderate or severe. Of these 76 children (with 50 boys), 67 were examined radiographically. The Cobb angle denotes the first radiographical examination after clinical diagnosis of a moderate or severe scoliosis.

No radiographic examination was performed in nine children; two died before an examination was possible, one was in too poor medical condition to submit to the procedure and the remaining six children were not referred to radiographical examinations immediately for unknown reasons. Further clinical examinations of these children graded their scoliosis type as mild so they were not examined radiographically. The proportion of children with scoliosis increased with GMFCS level. Almost all children with curves >20° were at GMFCS levels IV and V (Figure 9).

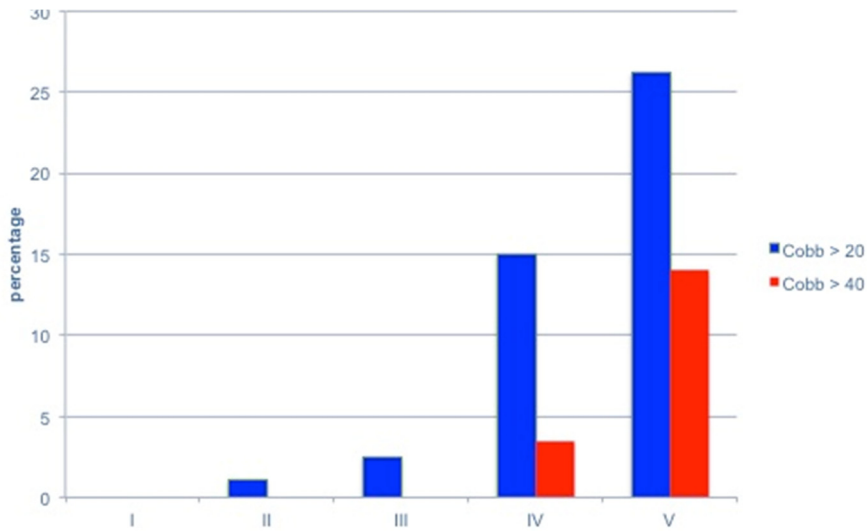


Figure 9.
Scoliosis in relation to GMFCS levels. Distribution of scoliosis (%) according to the clinical examination and first radiographical examination. Children with a Cobb angle of >40° are included in the group with a Cobb angle of > 20°.

The proportion of children with scoliosis varied between CP subtypes. No child with spastic unilateral CP, three of 75 children with ataxic CP, 38 of 244 with spastic bilateral CP and 10 of the 66 children with dyskinetic CP had a curve >20°. In total 18 children had surgery for scoliosis. The median age at surgery was 13 years (range 8-17). The mean preoperative Cobb angle was 69° (median 67°; range 40°-95°). Hip dislocation was present in nine children in this group. All were born in 1990-91 and had a moderate or severe scoliosis with Cobb angles of 70°-102°.

Kaplan-Meier survival estimations illustrated that a moderate or severe form of scoliosis was mostly diagnosed after 8 years of age, and that the risk increased with age and GMFCS level. Children in GMFCS levels I and I had almost no risk of developing scoliosis, but children in GMFCS levels IV and V had an approximately a 50% risk of having moderate or severe scoliosis at 18 years of age (Figure 10).

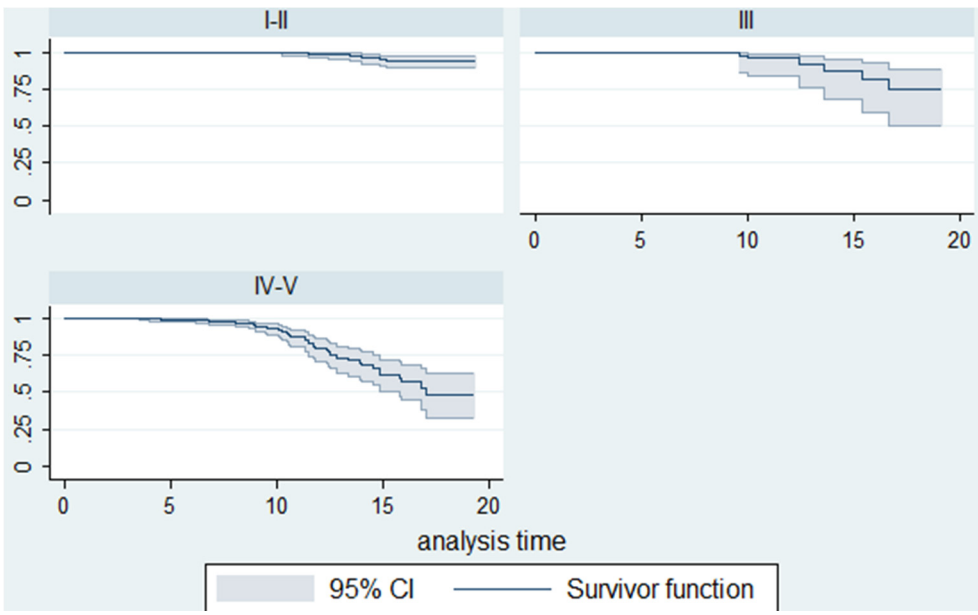


Figure 10.
Kaplan -Meier illustration of the survival with 95% CIs showing the risk of having moderate or severe scoliosis diagnosed at different ages and GMFCS levels.

Linear regression analyses showed that the only risk factor that influenced the magnitude of the Cobb angle at the first radiographical examination was the GMFCS level (p= 0.004); Table 4).

Table 4.
Linear regression analyzes of risk factors influencing the magnitude of Cobb angle.

	Coefficient	95% CI	P
GMFCS	8.09	2.78-13.39	0.004
Spastic bilateral CP	-9.17	-34.70 to 16.37	0.471
Ataxic CP	-12.67	-58.01 to 32.68	0.574
Dyskinetic CP	-21.11	-49.28 to- 7.05	0.137
Age	-0.20	-2.04 to 1.65	0.830

Cox regression analysis showed that a high GMFCS level was the most important factor for developing moderate or severe scoliosis (Table 5).

Table 5.
Cox regression analysis of the risk ratio (i.e. hazard ratio) for developing clinically moderate or severe scoliosis when considering the GMFCS level and CP subtype.

GMFCS/CP-type	Risk ratio	95% CI	p
II	2.70	0.68-10.65	0.156
III	6.04	1.52-23.99	0.011
IV	14.94	4.47-49.95	<0.001
V	34.99	10.74-113.98	<0.001
Spastic bilateral CP	0.85	0.27-2.71	0.787
Ataxic CP	0.79	0.17-3.35	0.749
Dyskinetic CP	0.53	0.14-1.92	0.330
Unclassified type	0.52	0.09-3.11	0.473

Reliability and validity of spinal assessment (*Study III*)

Twenty-eight children (14 girls) participated in the study, with a median age of 12 years (range 6-16) and GMFCS II-V.

There was an excellent interrater reliability for both the clinical examination (weighted kappa= 0.96) and scoliometer measurements (weighted kappa = 0.86).

The clinical examination showed a sensitivity of 75% (95% CI, 19.4-99.4%), a specificity of 99.8% (95% CI, 78.9-99.9%) and an AUC of 0.85 (95% CI, 0.61-1.00). The positive LR was 18.0 and the negative LR was 0.3.

The scoliometer measurement showed a sensitivity of 50% (95% CI, 6.8-93.2%), a specificity of 91.7% (95% CI, 73.0-99.9%) and AUC of 0.71 (95% CI, 0.42-0.99). The positive LR was 6.0 and the negative LR was 0.5.

Psychometric evaluation of PPAS (*Study IV*)

Twenty-nine children with CP (15 boys), participated in the study, with a median age of 12 years (range 6-16) with GMFCS II -V.

The PPAS showed excellent interrater reliability for three independent raters with weighted kappa values of 0.77-0.99 (95% CI, 0.60-1.0).

There was a high internal consistency for the PPAS for all items where Cronbach's alpha if an item was deleted was 0.95-0.96 with a 95% CI of 0.90-0.98 for all items. The corrected-item total correlation was 0.55 -0.91 (95% CI, 0.20-0.95).

The PPAS showed construct validity based on the ability of the assessment tool to distinguish between known groups ($p<0.01$) represented by GMFCS levels of II -V. Children at GMFCS level II had higher scores than children in GMFCS IV and V. The median score was higher with the children in supine or prone positions because these require less postural ability than standing or sitting positions.

The distribution of scores for all three raters varied between each GMFCS level at all four positions evaluated (Figure 11).

The PPAS differed for postural ability between individuals at different GMFCS levels and was able to identify postural asymmetries in children at all levels II-V.

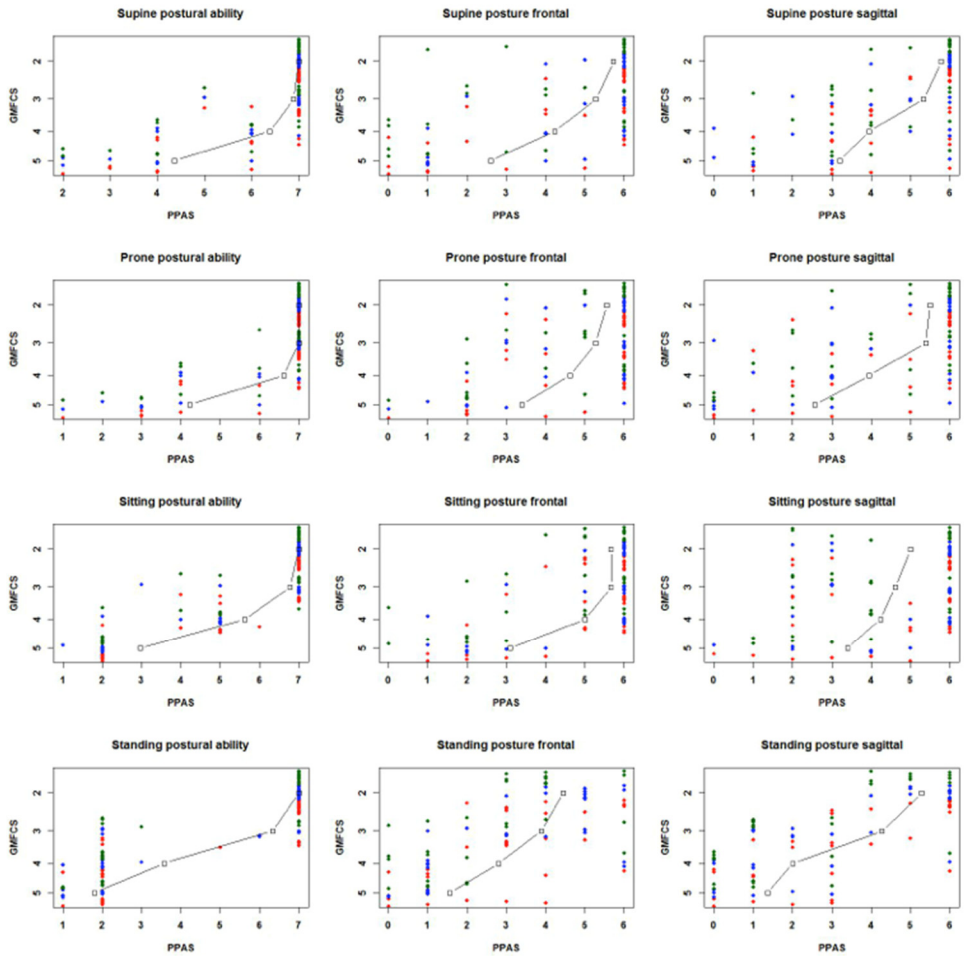


Figure 11.
Distribution of PPAS values at GMFCS levels II -V for all four postural positions. Observations are marked with a different color for each rater, red=rater A; blue=rater B,; green= rater C. The boxes connected with a line are the means of each GMFCS level.

Discussion

The aims of this thesis were to study the frequency of WS and scoliosis in children with CP and to evaluate clinical assessment tools used to screen for these conditions.

Studies ***I*** and ***II*** were cross-sectional analyses of the total population of children and adolescents with CP in the south of Sweden. They described the prevalence of WS and scoliosis in children with CP at all GMFCS levels and CP subtypes, given that the definitions of WS and scoliosis in this study were appropriate. Studies ***III*** and ***IV*** were psychometric evaluations of the clinical tools used to assess spinal deviations and postural asymmetries. These assessments are in clinical use, but have not been evaluated previously for children with CP. Although the children and adolescent comprised relatively small samples (28 and 29 individuals, respectively) the number of children considered to have scoliosis (***Study III***) was in accordance with the prevalence of scoliosis in a total population of children with CP (***Study II***) and therefore likely to be representative. In ***Study III*** the purpose was to evaluate the psychometric properties of the spinal screening procedure and to determine whether the method could identify individuals in need of further radiographical examination and thereby avoid unnecessary radiographs in individuals without significant scoliosis. This is why children with different levels of gross motor function were included without knowledge of their spinal condition. In ***Study IV***, the purpose was to evaluate the PPAS in children with CP.

Despite preventing hip dislocation, the incidence of WS and moderate or severe scoliosis were only partly reduced by the follow-up program. The possible development of contractures starting from the knees might explain some cases of WS and postural asymmetries that could be evaluated futher. Early and intensified treatment of contractures in the lower extremities, and of scoliosis with new surgical techniques that allow for further growth, might reduce the development of WS.

This thesis was partially based on registry studies. The efficacy of new treatment options can be analyzed by randomized controlled studies, but they need to be designed correctly with adequate power, and with one clinically important hypothesis and reliable endpoints. The ideal situation would be if randomized controlled studies and registry studies could complement each other and both could be used. Data from registry studies can be used to define endpoints, hypotheses and help to identify

confounding factors and randomized controlled studies can be used to evaluate specific treatments.

However, CP is a complex disorder affecting a heterogeneous population and studies need many years of follow-up to be able to evaluate which factor or factors might make registry studies favorable especially when in rare events and in large unselected populations.

Windswept hip deformity

The frequency of WS was 12% in the control group and 7% in the study group at 10 years and 18% and 9% at 20 years respectively. We used the same formula as constructed by Young et al. to define WS with the exception of including the measurement of adduction in terms of the ROM (77). Measurement of adduction is not performed in the CPUP. The reason for this is the low functional value of reduced adduction. Young et al. examined 103 children with spastic tetraplegic CP and found 52% WS and 25% whith hip dislocation. These prevalence figures are not comparable with ours because the study was made on a selected group of individuals with CP. Madigan and Wallace (41) studied a selected population of 272 institutionalized children with CP and found that 13% had WS. The diagnosis of WS was made on radiographs showing scoliosis, pelvic obliquity, and adduction of one hip and abduction of the other. All 36 children with WS had spastic bilateral CP, were nonambulant, and had scoliosis.

In the study by Young et al. (77) WS was associated with asymmetric tone and the side with the strongest tone in the adductors was more often dislocated or held in adduction. Their findings were consistent with those of Nwaobi et al. (96), who found disturbed electromyographic activity and an imbalance between adductor and abductor muscles in the hips of 13 tetraplegic children with WS. The activity was greater in the adductors. In our study we used the modified Ashworth scale (97) and found that only seven of the 18 children with WS had a higher muscle tone in the adductor muscles on the adducted side. Although the origin of the direction of the asymmetry creating sweeping of the legs is unclear, it is logical to consider that sustained asymmetric postures or asymmetric tone, in a combination with knee flexion contractures, might cause tilting of the legs to one side and start the development of WS. This can be looked upon as a body-shape distortion when the subject's knees fall to one side in a supine position and the pelvis drops in the opposite direction in search for a supporting surface. This creates a rotational force inducing an asymmetrical postural deformity that can influence the progression of hip migration and scoliosis (81). Several authors have investigated the direction of WS, pelvic obliquity, and scoliosis with the aim of finding an association with the side for

hip dislocation (76, 77, 81, 98-100). Letts et al. (76) found that the spinal curvature was convex away from the dislocated hip. The dislocation occurred on the high side of the pelvis and the WS was directed to the low side. These findings have been confirmed by some authors but not by others. Lonstein and Beck (19) studied 464 radiographs of subjects in a sitting position to analyze pelvic obliquity in dependent and independent sitters. They could not determine any association between the side of hip displacement, direction of pelvic obliquity, WS, or scoliosis. This might have been because the radiographic examinations were made with the subjects in a sitting position where the hips are flexed. In such postures, reduced abduction will cause a rotation of the pelvis and not an obliquity, while reduced hip flexion on one side causes pelvic obliquity which induces a rotation in the coronar plane and creates a secondary scoliosis. In the study, subjects with hip displacement and a MP value of >60 % were included, which is not comparable to complete dislocation. Porter (81) in a cross-sectional study, analyzed 747 individuals with CP aged 6 to 80, all in GMFCS level V. The convexity of the spinal curve was more likely to be opposite to the direction of WS, similar to the findings of Letts et al. (76).

In our study, the hip surveillance program did not significantly reduce the frequency of WS, but for children with WS and hip dislocation the reduction was statistically significant. In the study group, eight of 139 subjects were treated with a varization osteotomy of the proximal femur to prevent hip dislocation. In three of these children, it reduced the ROM in abduction to an extent that made them fulfill the WS criteria according to our definition. The imbalance induced by a unilateral osteotomy has led to proposals to operate on the contralateral unaffected hip prophylactically to reduce the risk of secondary displacement and create symmetry (101). A study by Larsson et al. (33) showed that the contralateral side had a low risk of later displacement after unilateral varization osteotomy. Owers et al. treated 30 children with WS and hip displacement with bilateral femoral osteotomy and bilateral soft tissue release, to produce symmetry. The relative frequency of WS had not been reduced significantly at follow-up after a median of 3.2 years (101).

The same cohort from ***Study I*** was further analyzed at 20 years of age (unpublished data). If the three children who were operated with a varization osteotomy of the proximal femur were excluded the frequency of WS in the study group would have been reduced to 5% ($p=0.028$). In the control group, another four children developed WS. In the study group, another three developed WS. Even if WS develops before 10 years of age in most children, the risk continues up to 20 years of age, which suggests the need for a follow-up in adults.

In conclusion, WS started at an early age and was associated with the level of gross motor dysfunction. A hip prevention program and early treatment of contractures seems to have reduced the frequency of this condition, which starts from the lower extremities.

Scoliosis

Among the 666 children in this study, 116 (17%) had mild, and another 76 (11%) had moderate or severe scoliosis based on clinical examinations. The risk of developing at least moderate scoliosis increased with GMFCS level. In most children, the scoliosis was diagnosed after 8 years of age.

This thesis analyzed the prevalence of scoliosis in a total population of children with CP, 4-18 years of age. It is difficult to compare the prevalence of scoliosis in our study with other studies representing selected groups of children with different definitions of scoliosis (34, 41, 102). The reported prevalence range is 15-80% (34). It seems that clinical examinations do not underestimate the degree of scoliosis. Many studies have defined scoliosis as a Cobb angle of >10°, and different images in supine and upright positions have been used during radiography (34).

The reliability and validity of the clinical spinal examination methods used in this study were evaluated in ***Study III***. In screening procedures, there is an inevitable balance between the sensitivity and specificity of the methods used. The uncertainty of the sensitivity and validity of our screening methods and how we defined scoliosis in this study was the starting point for testing the psychometric properties in this third part of the thesis.

The risk of being diagnosed with a moderate or severe scoliosis increased with GMFCS level and age. Almost all children (44/45) with curves >20° were in GMFCS levels III-V and all 18 children that were subjected to spinal fusion were in GMFCS levels IV and V. Earlier studies on the prevalence of scoliosis were undertaken before the GMFCS classification was introduced, but the risk of developing scoliosis is also associated with neurological impairments and age, as described earlier (34, 38, 43). The statistical analysis showed here that the GMFCS level was a better predictor than the CP subtype when estimating the risk of developing scoliosis in an individual child. The high frequency of children with spastic bilateral or dyskinetic CP and scoliosis is explained by the high proportion of GMFCS levels IV and V in those CP subtypes. The low prevalence of scoliosis in children with spastic unilateral CP might be explained by the high frequency of children in GMFCS levels I and II in that CP subtype. GMFCS is a reliable and valid measure to use and the GMFCS level remains relatively stable during maturation with time (15, 16).

Because children in GMFCS levels I and II have almost the same risk for developing scoliosis as for adolescent idiopathic scoliosis in normally developing children, similar screening procedures can be used, whereas children in GMFCS levels III -V need to be tracked with regular spinal examinations from early childhood and even into adulthood.

All nine children with hip dislocation in this population had moderate or severe scoliosis. Even if hip dislocation has been prevented by the CPUP, about 20% of the children aged 8-14 years and in GMFCS levels III-V had a moderate or severe scoliosis (www.CPUP.se/Årsrapport 2014). Children born in 1990-91 are followed in the CPUP but are not participating in the hip surveillance program. At 16 years of age, this population has a higher prevalence of scoliosis than did the population born in 1992-1997 (at 16 years) who are participating in the hip surveillance program. Although it appears that the frequency of scoliosis has been reduced in children following the hip surveillance program, other factors besides hip dislocation influence the prevalence of scoliosis. Better treatment of spasticity and contractures are possible explanations for this.

In conclusion, this study showed that GMFCS level and age were important factors for deciding a follow-up spinal evaluation. For children in GMFCS levels III-V regular spinal examinations in adulthood are recommended. Hip surveillance in the CPUP could only partially reduce the incidence of scoliosis.

Reliability and validity of spinal assessment

The purpose of the study was to evaluate whether the screening method used in CPUP was able to identify those in need of further radiographic investigations. The specificity of the clinical assessment was high (99.8%) indicating that no unnecessary referrals for radiographic examination were performed.

The clinical spinal assessment method used to screen for scoliosis in CPUP had excellent interrater reliability and a high concurrent validity when compared with radiographic Cobb angle measurement. When used in children with CP the scoliometer measurement was almost as reliable and valid but had no extra advantage. The clinical spinal assessment method seems appropriate as a screening tool to identify scoliosis that needs further evaluation by radiographic examination. In the CPUP, the results from spinal clinical examinations indicate who needs further evaluation by radiographic examination. The scoliometer or the clinical examination methods, to our knowledge, are not evaluated as screening tools for neuromuscular scoliosis. In children with CP, the scoliometer was more difficult to use, because some children had difficulties bending forward due to their limited hip flexion or when using an intrathecal baclofen pump.

Routine screening for adolescent idiopathic scoliosis has been questioned in terms of its cost benefit, but it is accepted and is a widely used method for the early detection of spinal curvatures (103). Pro-screening supporters state that early detection might change the natural history of this condition and reduce mortality (104). The

opponents argue that the cost outweighs the benefits and that it results in an unacceptable number of false positive findings. In a review by an expert panel (105), the scoliometer was recommended as being currently best screening tool. A referral for subjects with a value range of 5-7° can be recommended according to the reviewers. The optimal cutoff point for referral when using the scoliometer in school screening has been difficult to determine (103). The balance between too many false negative findings versus too many false positive results must be considered. Bunnell (106), who introduced the scoliometer in screening, first recommended 5° but later suggested 7° as sufficient, to avoid over referral. This is why 7° was used as cutoff point in the present study. If 5° had been chosen as the cutoff, twice as many children would have been referred for radiographic examinations. In only one case, it would have identified a child who had a Cobb angle of 23°. In all other cases the Cobb angles were <15°. In this study, the Cobb angle was used for defining scoliosis on the radiographic examination. We chose 20° as the cutoff because the usual definition of scoliosis as a Cobb angle of >10° would have resulted in too many insignificant curvatures. When defining scoliosis according to the Cobb angle, it is worth mentioning that both the interrater reliability and the intrarater variability values are approximately 5° (52, 53). In this study, 14 children who were regarded to have no or mild scoliosis had Cobb angles of 10-19°. If a Cobb angle of >10° was used as a valid cutoff for scoliosis these children with no or mild scoliosis would be regarded as having false negative findings. Several of these children with CP have reduced postural ability making it more likely for them to present with a small postural curve rather than structural scoliosis at radiographic examinations. For instance children with CP usually have a longer sweeping C-shaped scoliosis with a Cobb angle of 20°, and this differs from the shorter S-shaped curvatures found in adolescent idiopathic scoliosis.

The purpose of screening for adolescent idiopathic scoliosis is to detect it in time to start bracing and to reduce the need for surgery. The prevalence of adolescent idiopathic scoliosis is 2-4%. Among these approximately 8-9% will be treated with braces and only about 0.1% will be treated by surgery, depending on the indications used for treatment at different locations (105). Brace treatment is shown to be effective (107), unlike in children with CP where there is no evidence that it stops progression (57, 58). The reason for screening for neuromuscular scoliosis is to detect progressive scoliosis in time for surgery. Larger curves are often less flexible and the result of spinal surgery is related to the curve magnitude in general (50). The screening procedure used in CPUP even acts as a tool to detect postural asymmetries because asymmetric muscle tone, weakness and lack of stability might induce a deterioration in antigravity control that can increase the development of contractures, WS, scoliosis, and hip dislocation (78).

The psychometric properties of the scoliometer in evaluating adolescent idiopathic scoliosis have been studied before (108-110), but to our knowledge not during

screening procedures for neuromuscular scoliosis. However, the aim of this study was not to compare the clinical spinal assessment method with that using a scoliometer. The validity of both procedures was evaluated and compared with the Cobb angle measurements. The interrater reliability of the methods was tested and was excellent for both methods. In conclusion, the clinical spinal assessment method seems to be an appropriate screening method for scoliosis in children with CP. The validity was high when correlated with the Cobb angle measurement. The use of a scoliometer had no extra advantage.

Psychometric evaluation of PPAS

The PPAS showed high psychometric properties for children and adolescents with CP. The interrater reliability, construct validity, and internal consistency were all high and the results were similar to a previous study on adults (3). In that study on adults, the ratings were based on photographs and video recordings and the three raters were experienced physiotherapists who participated in the development of the PPAS. In the present study, the ratings were based on clinical examinations by three independent raters, but only one of them had experience of using PPAS. The interrater reliability was excellent for all three raters. We chose to perform the ratings at the same occasion, but the ratings were noted independently. Compliance among the children examined could have differed if the ratings were performed on different occasions.

Even if CP is a nonprogressive injury to the brain, secondary musculoskeletal abnormalities can start developing at an early age and continue during life (38, 49, 111, 112). The PPAS has been able to identify problems of posture and postural ability in adults, and this study has shown that it is also appropriate to use for children. The ability of the PPAS to identify postural deficits and asymmetries at an early age could thereby initiate early appropriate interventions, such as adaptive seating or standing, or noctural support (113-115). Asymmetric postures and the time spent in different locked positions might increase the risk of tissue adaptations that could lead to the development of contractures and progressive deformities (80, 82, 87). A study by Rodby-Bousquet et al. (78) showed that postural asymmetries were associated with scoliosis, hip dislocation, knee and hip contractures and an inability to change position. That study showed that there is an association between posture and limited ROM, but the question still to be answered is whether contractures are caused by the asymmetric posture or whether the limited ROM causes postural asymmetries.

Hip dislocation, WS, and scoliosis are associated with each other (35, 44) and a hip surveillance program might reduce their frequencies (26). Even if hip dislocation could be prevented, the frequency of scoliosis is still substantial among children at

lower levels of gross motor function. Other factors than the mechanics of a dislocated hip must influence the development of progressive scoliosis and pelvic obliquity (98).

Given that all these deformities are associated with postural asymmetries, it is important to include both an assessment of the ROM and posture in the follow up for children and adolesecents with CP from an early age and throughout life. The PPAS identified postural asymmetries in children at GMFCS levels II-V. Children at GMFCS level II can walk independently and have the highest level of PPAS concerning their ability to move into or out of position. An expected ceiling effect was seen in postural ability for these children. The PPAS is primarily designed for use with individuals at a lower level of gross motor function. However, it can be used to detect asymmetries in children and adults at all GMFCS levels.

The results from the assessments give indications for the potential need for postural support, and where it needs to be applied in attempting to prevent musculoskeletal deformities and improve function

The PPAS is simple to use and requires only a plinth and a scoring sheet. It takes about 10 minutes to complete in a clinical setting, but it is recommended to include some training and guidelines of what to be aware of in this setting. For instance, the internal consistency showed slightly lower values for subjects in a sitting posture in the sagittal view. This might be explained by the difficulty to assess whether the hips are flexed to 90° depending on the position of the pelvis and the height of the plinth. The height needs to be adjusted or foot support provided during any assessment of seated posture.

In conclusion the PPAS can be used in children and adolescents with CP to allow early detection of postural deficits and asymmetries. This can provide information about what types of postural support need to be applied.

Limitations

In ***Study I***, we did not analyze the number of children who had scoliosis without having WS. The frequencies of WS and scoliosis depend on the definitions being used. The statistically nonsignificant change in WS between the control and study groups may be because too few observations might miss an effect that actually exists (Type II error).

In ***Study II***, a limitation might be our definition of scoliosis. In CPUP, only children with moderate or severe clinical scoliosis according to the physiotherapist's examination, are referred to radiographical examination. Only a Cobb angle of >20° was regarded as scoliosis. Curves of <20° are usually regarded as clinically

nonsignificant and require no treatment besides a follow up to make sure there is no progression. Some children with a Cobb angle of >20° could have been classified as having only mild scoliosis and not examined radiographically. This was the reason for initiating ***Study III***, where we found the clinical examination method to have an appropriate sensitivity to identify those children with a >20° Cobb angle. The distribution of ages varied and some children were followed for a shorter time period and might not yet have developed scoliosis. This could have influenced the prevalence rates, although the Kaplan -Meier analysis considers that factor in the estimation. The study was cross-sectional, but had both prospective and retrospective aspects. Children were followed until the developing of a moderate scoliosis and the characteristics of these children were analyzed at that time.

In ***Study III***, only four of the 28 participating children had moderate or severe scoliosis. The study did not analyze the differences between moderate and severe scoliosis or between no or mild scoliosis. This was because of the cutoff points used, and the purpose of the study was to determine whether the method could safely select those in need for further investigation. The study did not attempt to predict the potential results if different cutoff values had been used for the Cobb angle and scoliometer measurements.

In ***Study IV***, one of the raters was experienced and had participated in the development of the PPAS. If all three examiners had been inexperienced, the results could have differed.

Conclusions

- Participation in the hip surveillance program of the CPUP significantly reduced the incidence of WS starting in the lower extremities, if the hips defined as having WS after femoral varization osteotomy were excluded. The frequency of WS at 10 years of age was 7% in the study group and 12% in the control group (nonsignificant). The incidence of WS increased even after 10 years of age, and at 20 years of age 9% of subjects in the study group and 18% in the control group had this syndrome. Because the risk of developing WS continues after skeletal maturity has been attained, there is a need for continuous follow-up in adulthood. In three of nine children in the study group, WS developed after femoral varus osteotomy done to prevent the hip from dislocation. WS was only seen in children at GMFCS levels III-V.
- The prevalence of mild scoliosis was 17%. The prevalence of moderate or severe scoliosis was 11%. The risk of developing scoliosis increased with GMFCS level and age. Children in GMFCS levels I and II had a low risk of developing scoliosis. Children and adolescents in GMFCS levels IV and V had a 50% risk of having moderate or severe scoliosis at 18 years of age. All nine children with a hip dislocation had a moderate or severe scoliosis. All children operated on for scoliosis were in GMFCS levels IV or V. The CP subtype was not a specific riskfactor for scoliosis because of different proportions of GMFCS levels in the subtypes studied. Follow-up programs for scoliosis should be based on the child's GMFCS level, age, Cobb angle, and the rate of progression.
- The clinical spinal assessments used to screen for scoliosis in children with CP had excellent interrater reliability and high concurrent validity when compared with the Cobb angle measurements. The clinical spinal assessments had slightly better psychometric properties than did the scoliometer measurement when used for children with CP. The clinical spinal assessment is appropriate for screening to identify spinal curvatures that need further investigation by radiographical examinations. It might be of importance that the clinical and radiographical examinations are both performed with the subject in a sitting position to ensure agreement between the methods, and to minimize measurement errors such as leg length discrepancies or contractures of the hip, knee and/or ankle.

- The PPAS had excellent interrater reliability, high internal consistency and good construct validity when evaluated for children with CP, as seen previously for adults. As expected, children in GMFCS level II had better postural ability than children in GMFCS levels IV and V. The PPAS could identify postural deficits and asymmetries at an early age in all GMFCS levels and could provide information on what type of postural support would be needed to prevent secondary musculoskeletal deformities and improve function. Therefore, it is recommended to assess both ROM and posture in addition to the clinical and radiographic follow up of hips and spine in children with CP from an early age and to continue this throughout adulthood to minimize deformities and pain, improve function, and ultimately optimize their quality of life.

Future aspects

This thesis analyzed the impact factor of a hip surveillance program on the prevalence of WS and scoliosis in children with CP. Two clinical examination methods for scoliosis and postural asymmetries and deformities were evaluated. These findings raise questions and ideas for further research.

Windswept hip deformity

Could WS be reduced by an early special spine surveillance program that identifies and treats scoliosis?

How does WS develop in adult years?

Scoliosis

What is the progression of scoliosis in adult years?

Could early treatment of scoliosis, with new techniques that allow for further growth, reduce the development of pelvic obliquity and WS?

Clinical examination of the spine to screen for scoliosis

What are the intrarater and interrater reliabilities of the clinical spinal examination method in known cases of scoliosis?

PPAS

If PPAS is used for children with CP, what are the consequences of an early identification of deformities?

It is important that all possible interventions and efforts that are made are also followed up and studied to evaluate whether if they improve the quality of life and participation in social activities as well as focusing on physical functions.

Acknowledgments and Grants

It is my honor and pleasure to thank all people who made this thesis possible.

I especially thank **all the children and adolescents with cerebral palsy who participated.** I learned a lot from you and hope that the knowledge gained from this thesis will be of some help. I admire your patience in being my patients.

Gunnar Hägglund, my scientific as well as clinical mentor and supervisor. The ”founder” of the iliopsoas muscle and the bridge between the hip and spine in this project. Your wisdom guided me every time I flexed my hips and sat down to write my thesis. It is impossible to overstate my gratitude to you. I truly appreciate your inspiration, insightfulness and determination in research. Thanks for great assistance and direct direction in writing my papers. Sometimes I wish I was you and when you’re not there I try to impersonate you. The impact factor when I mimic you has not yet been evaluated. Thank you for letting me participate in the CPUP. This project could have been called GHUP since all I know about CP and the advantages and skills to manoveur a follow up program eminates from you. You gave me the opportunity to work at the Pediatric Orthopedic Unit in Lund and you have always had the ability and fantastic capacity to give instant feedback and support whenever needed. It is a true pleasure working with you. You are a true long distance runner that never give up and inspire us all to Zlatanera.

Henrik Lauge-Pedersen, Co-tutor and Head of the Pediatric Orthopedic Unit. Harley Pedersen. Hvad gør vi nu lille du? Thanks for guidance and clinical support over the years. Thank you for helping me complete this thesis and to let me be a bit danish sometimes. My friend and boss. It works!

Elisabet Rodby-Bousquet, Co-tutor. Impressive energy. An essentiel vitamin for the metabolism of a project (invaluable input). Thanks for all support and shared knowledge. You impersonate the expression 24-7. You are like the enzyme who catalyzed the chain reaction which led to my final thesis. You tought me to move in and out of position and to keep my head in midline while working on my thesis.

My sincere thanks to:

All members of the pediatric orthopedic team in Lund-Malmö for showing me all the tricks at work, in the saddle and on and off the course.

A**ll my colleagues on ”plan 8”** for all encouragement and great discussions about life, work, research and everything in-between.

My former room mate and colleague Johan Nilsson who showed me the skills of orthopedic surgery and that the playlists on Spotify, as well as crucify pushups, might have a negative impact factor on research, but are of importance for our physical and mental health.

My present room mate and colleague Per-Ola Norrman. Thanks for helping me out in challenging clinical situations. Thanks for being a fantastic combination of friend, colleague and brother in-law. I am proud to be the uncle of your children. I think you appreciate that you have some hearing loss sometimes when I am present. Your research inspired me to finish mine.

Magnus Tägil, a friend and colleague and the best ”wine-red” professor in Lund. Thanks for the encouragement to finish my thesis and input and discussions about the academic theories in how to volontarily loose weight. Also, for the cheerful challenge in the Lund ”Social-knowledge and gossip update” competition.

Lena Westbom and all the staff at CPUP, for your support and expertise and important contributions when conducting the studies and for making this projekt possible.

Philippe Wagner and Tomaz Czuba, for your invaluable and instant support in interpreting data and statistical analysis.

My parents for all help, support and comfort throughout life when I’ve been in supine or prone positions. Sitting or standing. In frontal or sagital view. Whenever my body deviates from midline you fascilitate needed support. You have always supported me irrespectable of what self-initiated way of transportation I choose in life.

All my **brothers and sisters** for inspiration and guidance in life. We are so different, but so much alike. You always recieve my confessions. I am proud of our big family!

All friends from the past and present. My activities of daily living. My input in social life who bring me the statistics in social knowledge.

My **wife Sofia**, the power of love. The never ending support and understanding. Who has to compete with Häagen-Dazs, Lindt, Gruyere and Amarone, but these things never stand a chance against you. I am happy though that we share these moments together. My mental coach and PT. I certainly need you.

Fabian, Teodor and Disa. You’re the meaning in my life, you’re the inspiration. You give my life a purpose. I am happy that you inherited my sweet-tooth.

DJ, who has been the closest supporter sitting next to me when writing my thesis. Thanks for taking me out for a walk every fourth hour for an energy boost.

References

1. Rosenbaum P, Paneth N, Leviton A, Goldstein M, Bax M, Damiano D, et al. A report: the definition and classification of cerebral palsy April 2006. Developmental medicine and child neurology Supplement. 2007;109:8-14.
2. Reimers J. The stability of the hip in children. A radiological study of the results of muscle surgery in cerebral palsy. Acta Orthop Scand Suppl. 1980;184:1-100.
3. Rodby-Bousquet E, Agustsson A, Jonsdottir G, Czuba T, Johansson AC, Hagglund G. Interrater reliability and construct validity of the Posture and Postural Ability Scale in adults with cerebral palsy in supine, prone, sitting and standing positions. Clinical rehabilitation. 2014;28(1):82-90.
4. Lance JW. The control of muscle tone, reflexes, and movement: Robert Wartenberg Lecture. Neurology. 1980;30(12):1303-13.
5. Colver A, Fairhurst C, Pharoah PO. Cerebral palsy. Lancet. 2014;383(9924):1240-9.
6. Nordmark E, Hagglund G, Lagergren J. Cerebral palsy in southern Sweden I. Prevalence and clinical features. Acta paediatrica (Oslo, Norway : 1992). 2001;90(11):1271-6.
7. Westbom L, Bergstrand L, Wagner P, Nordmark E. Survival at 19 years of age in a total population of children and young people with cerebral palsy. Developmental medicine and child neurology. 2011;53(9):808-14.
8. Mac Keith RC, Mackenzie ICK, Polani PE. The Little Club Memorandum on Terminology and Classification of "Cerebral Palsy". Cerebral Palsy Bullentin. 1959;5:27-35.
9. Bax MC. Terminology and Classification of Cerebral Palsy. Developmental medicine and child neurology. 1964;6:295-7.
10. Mutch L, Alberman E, Hagberg B, Kodama K, Perat MV. Cerebral palsy epidemiology: where are we now and where are we going? Dev Med Child Neurol. 1992;34(6):547-51.
11. Hagberg B, Hagberg G, Olow I. The changing panorama of cerebral palsy in Sweden 1954-1970. I. Analysis of the general changes. Acta paediatrica Scandinavica. 1975;64(2):187-92.
12. Surveillance of cerebral palsy in Europe: a collaboration of cerebral palsy surveys and registers. Surveillance of Cerebral Palsy in Europe (SCPE). Developmental medicine and child neurology. 2000;42(12):816-24.

13. Palisano R, Rosenbaum P, Walter S, Russell D, Wood E, Galuppi B. Development and reliability of a system to classify gross motor function in children with cerebral palsy. Developmental medicine and child neurology. 1997;39(4):214-23.
14. Bodkin AW, Robinson C, Perales FP. Reliability and Validity of the Gross Motor Function Classification System for Cerebral Palsy. Pediatric Physical Therapy. 2003;15(4):247-52.
15. Imms C, Carlin J, Eliasson AC. Stability of caregiver-reported manual ability and gross motor function classifications of cerebral palsy. Developmental medicine and child neurology. 2010;52(2):153-9.
16. Palisano RJ, Cameron D, Rosenbaum PL, Walter SD, Russell D. Stability of the gross motor function classification system. Developmental medicine and child neurology. 2006;48(6):424-8.
17. Palisano RJ, Rosenbaum P, Bartlett D, Livingston MH. Content validity of the expanded and revised Gross Motor Function Classification System. Developmental medicine and child neurology. 2008;50(10):744-50.
18. Westbom L, Hagglund G, Nordmark E. Cerebral palsy in a total population of 4-11 year olds in southern Sweden. Prevalence and distribution according to different CP classification systems. BMC pediatrics. 2007;7:41.
19. Lonstein JE, Beck K. Hip dislocation and subluxation in cerebral palsy. Journal of pediatric orthopedics. 1986;6(5):521-6.
20. Bleck EE. The hip in cerebral palsy. The Orthopedic clinics of North America. 1980;11(1):79-104.
21. Hagglund G, Lauge-Pedersen H, Wagner P. Characteristics of children with hip displacement in cerebral palsy. BMC musculoskeletal disorders. 2007;8:101.
22. Robb JE, Hagglund G. Hip surveillance and management of the displaced hip in cerebral palsy. Journal of children's orthopaedics. 2013;7(5):407-13.
23. Larnert P, Risto O, Hagglund G, Wagner P. Hip displacement in relation to age and gross motor function in children with cerebral palsy. Journal of children's orthopaedics. 2014;8(2):129-34.
24. Soo B, Howard JJ, Boyd RN, Reid SM, Lanigan A, Wolfe R, et al. Hip displacement in cerebral palsy. The Journal of bone and joint surgery American volume. 2006;88(1):121-9.
25. Shore B, Spence D, Graham H. The role for hip surveillance in children with cerebral palsy. Current reviews in musculoskeletal medicine. 2012;5(2):126-34.
26. Hagglund G, Andersson S, Duppe H, Lauge-Pedersen H, Nordmark E, Westbom L. Prevention of dislocation of the hip in children with cerebral palsy. The first ten years of a population-based prevention programme. The Journal of bone and joint surgery British volume. 2005;87(1):95-101.
27. Hagglund G, Alriksson-Schmidt A, Lauge-Pedersen H, Rodby-Bousquet E, Wagner P, Westbom L. Prevention of dislocation of the hip in children with cerebral palsy: 20-year results of a population-based prevention programme. The bone & joint journal. 2014;96-b(11):1546-52.

28. Elkamil AI, Andersen GL, Hägglund G, Lamvik T, Skranes J, Vik T. Prevalence of hip dislocation among children with cerebral palsy in regions with and without a surveillance programme: a cross sectional study in Sweden and Norway. BMC musculoskeletal disorders. 2011;12:284.
29. Hagglund G, Andersson S, Duppe H, Lauge-Pedersen H, Nordmark E, Westbom L. Prevention of severe contractures might replace multilevel surgery in cerebral palsy: results of a population-based health care programme and new techniques to reduce spasticity. Journal of pediatric orthopedics Part B. 2005;14(4):269-73.
30. Hagglund G, Lauge-Pedersen H, Persson M. Radiographic threshold values for hip screening in cerebral palsy. Journal of children's orthopaedics. 2007;1(1):43-7.
31. Hermanson M, Hagglund G, Riad J, Wagner P. Head-shaft angle is a risk factor for hip displacement in children with cerebral palsy. Acta orthopaedica. 2014:1-4.
32. Martinsson C, Himmelmann K. Effect of weight-bearing in abduction and extension on hip stability in children with cerebral palsy. Pediatric physical therapy : the official publication of the Section on Pediatrics of the American Physical Therapy Association. 2011;23(2):150-7.
33. Larsson M, Hagglund G, Wagner P. Unilateral varus osteotomy of the proximal femur in children with cerebral palsy: a five-year follow-up of the development of both hips. Journal of children's orthopaedics. 2012;6(2):145-51.
34. Koop SE. Scoliosis in cerebral palsy. Developmental medicine and child neurology. 2009;51 Suppl 4:92-8.
35. Persson-Bunke M, Hagglund G, Lauge-Pedersen H, Wagner P, Westbom L. Scoliosis in a total population of children with cerebral palsy. Spine. 2012;37(12):E708-13.
36. Willner S, Uden A. A prospective prevalence study of scoliosis in Southern Sweden. Acta orthopaedica Scandinavica. 1982;53(2):233-7.
37. Ersberg A, Gerdhem P. Pre- and postoperative quality of life in patients treated for scoliosis. Acta orthopaedica. 2013;84(6):537-43.
38. Saito N, Ebara S, Ohotsuka K, Kumeta H, Takaoka K. Natural history of scoliosis in spastic cerebral palsy. Lancet. 1998;351(9117):1687-92.
39. A IT. Development and treatment of spinal deformity in patients with cerebral palsy. Indian journal of orthopaedics. 2010;44(2):148-58.
40. Kalen V, Conklin MM, Sherman FC. Untreated scoliosis in severe cerebral palsy. Journal of pediatric orthopedics. 1992;12(3):337-40.
41. Madigan RR, Wallace SL. Scoliosis in the institutionalized cerebral palsy population. Spine. 1981;6(6):583-90.
42. Lipton GE, Miller F, Dabney KW, Altiok H, Bachrach SJ. Factors predicting postoperative complications following spinal fusions in children with cerebral palsy. Journal of spinal disorders. 1999;12(3):197-205.
43. Majd ME, Muldowny DS, Holt RT. Natural history of scoliosis in the institutionalized adult cerebral palsy population. Spine. 1997;22(13):1461-6.

44. Persson-Bunke M, Hagglund G, Lauge-Pedersen H. Windswept hip deformity in children with cerebral palsy. Journal of pediatric orthopedics Part B. 2006;15(5):335-8.
45. Miller F. Spinal Deformity Secondary to Impaired Neurologic Control2007. 143-7 p.
46. Hodgkinson I, Berard C, Chotel F, Berard J. [Pelvic obliquity and scoliosis in non-ambulatory patients with cerebral palsy: a descriptive study of 234 patients over 15 years of age]. Revue de chirurgie orthopedique et reparatrice de l'appareil moteur. 2002;88(4):337-41.
47. Cooke PH, Cole WG, Carey RP. Dislocation of the hip in cerebral palsy. Natural history and predictability. The Journal of bone and joint surgery British volume. 1989;71(3):441-6.
48. Loeters MJ, Maathuis CG, Hadders-Algra M. Risk factors for emergence and progression of scoliosis in children with severe cerebral palsy: a systematic review. Developmental medicine and child neurology. 2010;52(7):605-11.
49. Thometz JG, Simon SR. Progression of scoliosis after skeletal maturity in institutionalized adults who have cerebral palsy. The Journal of bone and joint surgery American volume. 1988;70(9):1290-6.
50. Miyanji F, Slobogean GP, Samdani AF, Betz RR, Reilly CW, Slobogean BL, et al. Is larger scoliosis curve magnitude associated with increased perioperative health-care resource utilization?: a multicenter analysis of 325 adolescent idiopathic scoliosis curves. The Journal of bone and joint surgery American volume. 2012;94(9):809-13.
51. Cobb J. Outline for the study of scoliosis. In Instructional Course Lectures Rosemont I, editor: American Academy of Orthopaedic Surgeons 1948.
52. Pruijs JE, Hageman MA, Keessen W, van der Meer R, van Wieringen JC. Variation in Cobb angle measurements in scoliosis. Skeletal radiology. 1994;23(7):517-20.
53. Morrissy RT, Goldsmith GS, Hall EC, Kehl D, Cowie GH. Measurement of the Cobb angle on radiographs of patients who have scoliosis. Evaluation of intrinsic error. The Journal of bone and joint surgery American volume. 1990;72(3):320-7.
54. Driscoll SW, Skinner J. Musculoskeletal complications of neuromuscular disease in children. Physical medicine and rehabilitation clinics of North America. 2008;19(1):163-94, viii.
55. Lonstein JE, Akbarnia A. Operative treatment of spinal deformities in patients with cerebral palsy or mental retardation. An analysis of one hundred and seven cases. The Journal of bone and joint surgery American volume. 1983;65(1):43-55.
56. Bunnell WP, MacEwen GD. Non-operative treatment of scoliosis in cerebral palsy: preliminary report on the use of a plastic jacket. Developmental medicine and child neurology. 1977;19(1):45-9.
57. Olafsson Y, Saraste H, Al-Dabbagh Z. Brace treatment in neuromuscular spine deformity. Journal of pediatric orthopedics. 1999;19(3):376-9.
58. Miller A, Temple T, Miller F. Impact of orthoses on the rate of scoliosis progression in children with cerebral palsy. Journal of pediatric orthopedics. 1996;16(3):332-5.

59. Terjesen T, Lange JE, Steen H. Treatment of scoliosis with spinal bracing in quadriplegic cerebral palsy. Developmental medicine and child neurology. 2000;42(7):448-54.

60. Vekerdy Z. Management of seating posture of children with cerebral palsy by using thoracic-lumbar-sacral orthosis with non-rigid SIDO frame. Disability and rehabilitation. 2007;29(18):1434-41.

61. Letts M, Rathbone D, Yamashita T, Nichol B, Keeler A. Soft Boston orthosis in management of neuromuscular scoliosis: a preliminary report. Journal of pediatric orthopedics. 1992;12(4):470-4.

62. Leopando MT, Moussavi Z, Holbrow J, Chernick V, Pasterkamp H, Rempel G. Effect of a Soft Boston Orthosis on pulmonary mechanics in severe cerebral palsy. Pediatric pulmonology. 1999;28(1):53-8.

63. McCarthy JJ, D'Andrea LP, Betz RR, Clements DH. Scoliosis in the child with cerebral palsy. The Journal of the American Academy of Orthopaedic Surgeons. 2006;14(6):367-75.

64. Reames DL, Smith JS, Fu KM, Polly DW, Jr., Ames CP, Berven SH, et al. Complications in the surgical treatment of 19,360 cases of pediatric scoliosis: a review of the Scoliosis Research Society Morbidity and Mortality database. Spine. 2011;36(18):1484-91.

65. Jones-Quaidoo SM, Yang S, Arlet V. Surgical management of spinal deformities in cerebral palsy. A review. Journal of neurosurgery Spine. 2010;13(6):672-85.

66. Sitoula P, Holmes L, Jr., Sees J, Rogers K, Dabney K, Miller F. The Long-term Outcome of Early Spine Fusion for Scoliosis in Children with Cerebral Palsy. Journal of spinal disorders & techniques. 2014.

67. Watanabe K, Lenke LG, Daubs MD, Watanabe K, Bridwell KH, Stobbs G, et al. Is spine deformity surgery in patients with spastic cerebral palsy truly beneficial?: a patient/parent evaluation. Spine. 2009;34(20):2222-32.

68. Teli MG, Cinnella P, Vincitorio F, Lovi A, Grava G, Brayda-Bruno M. Spinal fusion with Cotrel-Dubousset instrumentation for neuropathic scoliosis in patients with cerebral palsy. Spine. 2006;31(14):E441-7.

69. Lonstein JE, Koop SE, Novachek TF, Perra JH. Results and complications after spinal fusion for neuromuscular scoliosis in cerebral palsy and static encephalopathy using luque galveston instrumentation: experience in 93 patients. Spine. 2012;37(7):583-91.

70. Larsson EL, Aaro SI, Normelli HC, Oberg BE. Long-term follow-up of functioning after spinal surgery in patients with neuromuscular scoliosis. Spine. 2005;30(19):2145-52.

71. McElroy MJ, Sponseller PD, Dattilo JR, Thompson GH, Akbarnia BA, Shah SA, et al. Growing rods for the treatment of scoliosis in children with cerebral palsy: a critical assessment. Spine. 2012;37(24):E1504-10.

72. Tsirikos AI, Chang WN, Dabney KW, Miller F. Comparison of parents' and caregivers' satisfaction after spinal fusion in children with cerebral palsy. Journal of pediatric orthopedics. 2004;24(1):54-8.

73. Tsirikos AI, Lipton G, Chang WN, Dabney KW, Miller F. Surgical correction of scoliosis in pediatric patients with cerebral palsy using the unit rod instrumentation. Spine. 2008;33(10):1133-40.

74. Bohtz C, Meyer-Heim A, Min K. Changes in health-related quality of life after spinal fusion and scoliosis correction in patients with cerebral palsy. Journal of pediatric orthopedics. 2011;31(6):668-73.

75. Larsson EL, Aaro S, Oberg B. Activities and functional assessment 1 year after spinal fusion for paralytic scoliosis. European spine journal : official publication of the European Spine Society, the European Spinal Deformity Society, and the European Section of the Cervical Spine Research Society. 1999;8(2):100-9.

76. Letts M, Shapiro L, Mulder K, Klassen O. The windblown hip syndrome in total body cerebral palsy. Journal of pediatric orthopedics. 1984;4(1):55-62.

77. Young NL, Wright JG, Lam TP, Rajaratnam K, Stephens D, Wedge JH. Windswept Hip Deformity in Spastic Quadriplegic Cerebral Palsy. Pediatric Physical Therapy. 1998;10(3):94-100.

78. Rodby-Bousquet E, Czuba T, Hagglund G, Westbom L. Postural asymmetries in young adults with cerebral palsy. Developmental medicine and child neurology. 2013;55(11):1009-15.

79. Bouisset S, Do MC. Posture, dynamic stability, and voluntary movement. Neurophysiol Clin. 2008;38(6):345-62.

80. Brown K. Positional deformity in children with cerebral palsy. Physiotherapy theory and practice. 1985;1(1):37-41.

81. Porter D, Michael S, Kirkwood C. Patterns of postural deformity in non-ambulant people with cerebral palsy: what is the relationship between the direction of scoliosis, direction of pelvic obliquity, direction of windswept hip deformity and side of hip dislocation? Clinical rehabilitation. 2007;21(12):1087-96.

82. Fulford FE, Brown JK. Position as a cause of deformity in children with cerebral palsy. Developmental medicine and child neurology. 1976;18(3):305-14.

83. Renshaw TS, Green NE, Griffin PP, Root L. Cerebral Palsy: Ortopaedic Management. The Journal of bone and joint surgery American volume. 1995;77(10):1590-606

84. Pope PM. Severe and complex neurological disability : management of the physical condition. Edinburgh: Butterworth-Heinemann/Elsevier; 2007.

85. Frank JS, Earl M. Coordination of posture and movement. Physical therapy. 1990;70(12):855-63.

86. Massion J. Postural control systems in developmental perspective. Neurosci Biobehav Rev. 1998;22(4):465-72.

87. Porter D, Michael S, Kirkwood C. Is there a relationship between preferred posture and positioning in early life and the direction of subsequent asymmetrical postural deformity in non ambulant people with cerebral palsy? Child: care, health and development. 2008;34(5):635-41.

88. Bunnell WP. An objective criterion for scoliosis screening. The Journal of bone and joint surgery American volume. 1984;66(9):1381-7.

89. Pepe MS. The statistical evaluation of medical tests for classification and prediction. Oxford ; New York: Oxford University Press; 2003. xvi, 302 p. p.
90. Conger AJ. Integration and generalization of kappas for multiple raters. Psychological Bulletin. 1980;88(2):322-8.
91. Fleiss J, L. , editor. Statistical Methods for Rates and Proportions. Second ed1981.
92. DiCiccio TJ, Efron B. Bootstrap Confidence Intervals. Statistical Science. 1996;11(3):189-212.
93. Davison ACaH, D.V., editor. Bootstrap Methods and Their Application, Chapter 5. : Cambridge University Press; 1997.
94. Grimes DA, Schulz KF. Refining clinical diagnosis with likelihood ratios. Lancet. 2005;365(9469):1500-5.
95. Streiner DL, Norman GR. Health measurement scales : a practical guide to their development and use. Oxford: Oxford University Press; 2008.
96. Nwaobi OM, Sussman MD. Electromyographic and force patterns of cerebral palsy patients with windblown hip deformity. Journal of pediatric orthopedics. 1990;10(3):382-8.
97. Bohannon RW, Smith MB. Interrater reliability of a modified Ashworth scale of muscle spasticity. Physical therapy. 1987;67(2):206-7.
98. Pritchett JW. Treated and untreated unstable hips in severe cerebral palsy. Developmental medicine and child neurology. 1990;32(1):3-6.
99. Abel MF, Blanco JS, Pavlovich L, Damiano DL. Asymmetric hip deformity and subluxation in cerebral palsy: an analysis of surgical treatment. Journal of pediatric orthopedics. 1999;19(4):479-85.
100. Black BE, Griffin PP. The cerebral palsied hip. Clinical orthopaedics and related research. 1997(338):42-51.
101. Owers KL, Pyman J, Gargan MF, Witherow PJ, Portinaro NM. Bilateral hip surgery in severe cerebral palsy a preliminary review. The Journal of bone and joint surgery British volume. 2001;83(8):1161-7.
102. Balmer GA, MacEwen GD. The incidence and treatment of scoliosis in cerebral palsy. The Journal of bone and joint surgery British volume. 1970;52(1):134-7.
103. Huang SC. Cut-off point of the Scoliometer in school scoliosis screening. Spine. 1997;22(17):1985-9.
104. Torell G, Nordwall A, Nachemson A. The changing pattern of scoliosis treatment due to effective screening. The Journal of bone and joint surgery American volume. 1981;63(3):337-41.
105. Labelle H, Richards SB, De Kleuver M, Grivas TB, Luk KD, Wong HK, et al. Screening for adolescent idiopathic scoliosis: an information statement by the scoliosis research society international task force. Scoliosis. 2013;8:17.
106. Bunnell WP. Outcome of spinal screening. Spine. 1993;18(12):1572-80.

107. Weinstein SL, Dolan LA, Wright JG, Dobbs MB. Effects of bracing in adolescents with idiopathic scoliosis. The New England journal of medicine. 2013;369(16):1512-21.

108. Coelho DM, Bonagamba GH, Oliveira AS. Scoliometer measurements of patients with idiopathic scoliosis. Brazilian journal of physical therapy. 2013;17(2):179-84.

109. Bonagamba GH, Coelho DM, Oliveira AS. Inter and intra-rater reliability of the scoliometer. Revista brasileira de fisioterapia (Sao Carlos (Sao Paulo, Brazil)). 2010;14(5):432-8.

110. Amendt LE, Ause-Ellias KL, Eybers JL, Wadsworth CT, Nielsen DH, Weinstein SL. Validity and reliability testing of the Scoliometer. Physical therapy. 1990;70(2):108-17.

111. Morrell DS, Pearson JM, Sauser DD. Progressive bone and joint abnormalities of the spine and lower extremities in cerebral palsy. Radiographics : a review publication of the Radiological Society of North America, Inc. 2002;22(2):257-68.

112. Kerr Graham H, Selber P. Musculoskeletal aspects of cerebral palsy. The Journal of bone and joint surgery British volume. 2003;85(2):157-66.

113. Pountney T, Mandy A, Green E, Gard P. Management of hip dislocation with postural management. Child: care, health and development. 2002;28(2):179-85.

114. Pope PM, Bowes CE, Booth E. Postural control in sitting the SAM system: evaluation of use over three years. Developmental medicine and child neurology. 1994;36(3):241-52.

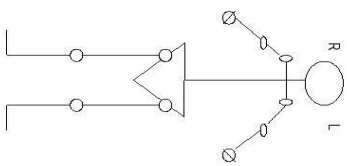
115. Rodby-Bousquet E, Hägglund G. Sitting and standing performance in a total population of children with cerebral palsy: a cross-sectional study. BMC musculoskeletal disorders. 2010;11(1):131.

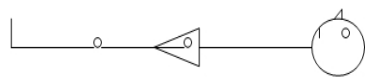
Appendix

Posture and Postural Ability Scale, Client _____ Date_____

LEVEL OF POSTURAL ABILITY IN SUPINE (*client on a mat, plinth or bed)*

Quantity		
Level 1	Unplaceable in an aligned supine posture	
Level 2	Placeable in an aligned supine posture but needs support	
Level 3	Able to maintain supine when placed but cannot move	
Level 4	Able to initiate flexion of trunk (stabilise trunk to lift head or knees)	
Level 5	Able to transfer weight laterally and regain posture (roll to the side)	
Level 6	Able to move out of supine position (i.e roll into prone)	
Level 7	Able to move into and out of supine position (i.e into sitting and back)	





QUALITY OF POSTURE IN SUPINE

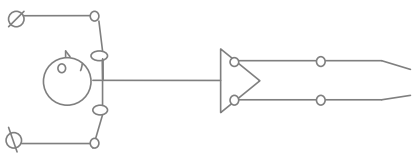
Quality, frontal (score 1=yes, 0=no)	
Head midline	
Trunk symmetrical	
Pelvis neutral	
Legs separated and straight relative to pelvis	
Arms resting by side	
Weight evenly distributed	
Total score	

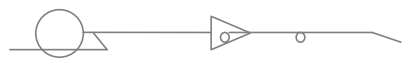
Quality, sagital (score 1=yes, 0=no)	
Head midline (flat pillow)	
Trunk in neutral position	
Pelvis neutral	
Legs straight, hips and knees extended	
Feet resting in normal position	
Weight evenly distributed (through shoulder girdle and pelvis)	
Total score	

Posture and Postural Ability Scale, Client _____ Date_____

LEVEL OF POSTURAL ABILITY IN PRONE *(client on a mat, plinth or bed)*

Quantity		
Level 1	Unplaceable in an aligned prone posture	
Level 2	Placeable in an aligned prone posture but needs support	
Level 3	Able to maintain prone when placed but cannot move	
Level 4	Able to initiate extension of trunk (lift and move head freely)	
Level 5	Able to transfer weight laterally and regain posture (roll to the side)	
Level 6	Able to move out of prone position (i.e roll into supine)	
Level 7	Able to move into and out of prone position (i.e into crawl and back)	





QUALITY OF POSTURE IN PRONE

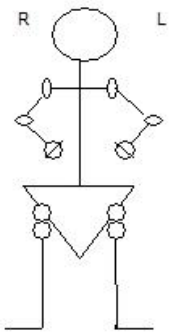
Quality, frontal (score 1=yes, 0=no)	
Head to one side	
Trunk symmetrical	
Pelvis neutral	
Legs separated and straight relative to pelvis	
Arms resting (elevated to mid position, upper arms resting & approx. 90° elbow)	
Weight evenly distributed	
Total score	

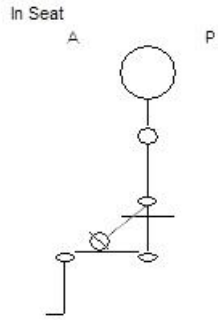
Quality, sagital (score 1=yes, 0=no)	
Trunk in neutral position	
Pelvis neutral	
Hips extended (feet off the end of table)	
Knees extended (feet off the end of table)	
Arms resting (elevated to mid position, upper arm resting & approx. 90°elbow)	
Weight evenly distributed (through shoulder girdle and pelvis)	
Total score	

Posture and Postural Ability Scale, Client _____ Date_____

LEVEL OF POSTURAL ABILITY IN SITTING *(client placed in sitting on a box or over the edge of a plinth with feet supported)*

Quantity		
Level 1	Unplaceable in an aligned sitting posture	
Level 2	Placeable in an aligned sitting posture but needs support	
Level 3	Able to maintain sitting when placed but cannot move	
Level 4	Able to move trunk slightly forwards-backwards over base without arching spine	
Level 5	Able to transfer weight laterally and regain posture (from one buttock to the other)	
Level 6	Able to move out of sitting position (i.e transfer weight onto feet and lift bottom of seat)	
Level 7	Able to move into and out of sitting position (i.e into standing and back)	





QUALITY OF POSTURE IN SITTING

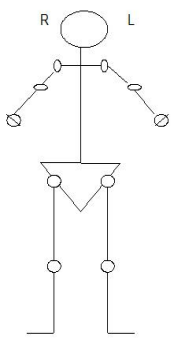
Quality, frontal (score 1=yes, 0=no)	
Head midline	
Trunk symmetrical	
Pelvis neutral	
Legs separated and straight relative to pelvis	
Arms resting by side	
Weight evenly distributed	
Total score	

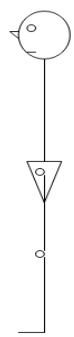
Quality, sagital (score 1=yes, 0=no)	
Head midline	
Trunk in neutral position	
Pelvis neutral	
Hips mid-position (90°)	
Knees mid-position (90°)	
Feet mid-position/flat on floor	
Total score	

Posture and Postural Ability Scale, Client _____ Date_____

LEVEL OF POSTURAL ABILITY IN STANDING

Quantity		
Level 1	Unplaceable in an aligned standing posture	
Level 2	Placeable in an aligned standing posture but needs support	
Level 3	Able to maintain standing when placed but cannot move	
Level 4	Able to move trunk slightly forwards-backwards over base without arching spine	
Level 5	Able to transfer weight laterally and regain posture (from one foot to the other)	
Level 6	Able to move out of standing position (i.e take a step forwards)	
Level 7	Able to move into and out of standing position (i.e take steps, walk)	





QUALITY OF POSTURE IN STANDING

Quality, frontal (score 1=yes, 0=no)	
Head midline	
Trunk symmetrical	
Pelvis neutral	
Legs separated and straight relative to pelvis	
Arms resting by side	
Weight evenly distributed (through both feet)	
Total score	

Quality, sagital (score 1=yes, 0=no)	
Head midline	
Trunk in neutral position	
Pelvis neutral	
Legs straight, hips & knees extended	
Feet mid-position/flat on floor	
Weight evenly distributed (through the feet)	
Total score	

Study 1

Windswept hip deformity in children with cerebral palsy

Måns Persson-Bunke, Gunnar Hägglund and Henrik Lauge-Pedersen

Windswept hip deformity describes an abduction and external rotation position of one hip with the opposite hip in adduction and internal rotation. Windswept hip deformity may occur in association with hip dislocation and scoliosis. We analysed the prevalence of this deformity in a total population of children with cerebral palsy, and the impact of hip prevention and early treatment of contractures on the prevalence and severity of windswept hip deformity. The frequency of windswept hip deformity was 12% in the control group and 7% in the study group, comprising children in the hip prevention programme. The children with this deformity in the study group had a lower frequency of scoliosis and none had hip dislocation. It thus seems that the hip prevention programme results in a decrease in the number of children with windswept hip deformity, and a decrease in the severity of the deformity. ***J Pediatr Orthop B* 15:335–338**

Journal of Pediatric Orthopaedics B 2006, **15**:335–338

Keywords: cerebral palsy, hip dislocation, scoliosis, windswept

Department of Orthopaedics, Lund University Hospital, Lund, Sweden

Correspondence and requests for reprints to Dr Gunnar Hägglund, PhD, Department of Orthopaedics, Lund University Hospital, S 221 85 Lund, Sweden
E-mail: Gunnar.Hagglund@med.lu.se, Mans.Persson-Bunke@med.lu.se

Introduction

Children with cerebral palsy (CP) often have an increased muscle tone, muscle weakness and muscle imbalance. This gives them an increased risk for developing muscle contracture, hip dislocation and scoliosis. Windswept hip deformity (WS) is a clinical manifestation among some children with CP. The term describes an abduction and external rotation position of one hip with the opposite hip in adduction and internal rotation [1]. WS is sometimes preceded by hip dislocation and sometimes by scoliosis. It is a severe problem that is difficult to treat, and that inhibits seating comfort and standing.

A CP register and health care programme aimed at preventing hip dislocation and severe contracture was initiated in southern Sweden in 1994 [2,3]. The total population of children with CP in the area, born in 1990 or later, is included. We have used this material to study:

1. The prevalence of WS in a total population of children with CP.
2. The impact of hip prevention, and early treatment of contractures on the prevalence and severity of WS.

Material and methods

The study area has a population of 1.3 million inhabitants. All children with CP in the area, born in 1990 or later, are included in the CP register [4]. Children born in 1992 or later are included in a hip prevention programme. The present study includes the children with CP born during 1990–1995. Only those who were born in the area or had moved into the area before 2 years of age and were still living in the area at 10 years of age and participating in the follow-up programme were included. A classification of subtype of CP was carried out according to Hagberg *et al.* [5]. The gross motor function was classified according to the Gross Motor Function Classification System (GMFCS) [6], a five-level age-related system in which level I is the least and level V the most affected.

The children born during 1990–1991 (not participating in the hip prevention programme) were regarded as the control group. The children born during 1992–1995 constituted the study group. We identified 207 children fulfilling the criteria for inclusion, 68 were born during 1990–1991 and 139 were born during 1992–1995 (Table 1).

The health care programme includes a standardized follow-up of the children's gross motor function, clinical findings and treatment. The child's local physiotherapist and occupational therapist measure and fill in a recording form twice a year until the age of 6 years and then once a year. The clinical findings include ROM measurements with a goniometer, measurement of muscle tone with the Ashworth scale [7] and measurement of scoliosis. The health care programme also includes a radiological follow-up of the hips in children born in 1992 or later. All children with signs of scoliosis were examined radiographically.

From the database, it is possible to follow each child's hip range of motion continuously. Children with bilateral CP and at least 50% difference in abduction, internal and/or external rotation between left and right hip were defined as WS. At least two consecutive measurements with this difference, with at least a 6-month interval, were required

1060-152X

Table 1 Number of children with cerebral palsy in the control group (born during 1990–1991) and the study group (born during 1992–1995)

	Born during 1990–1991	Born during 1992–1995	Total
Born in the area during 1990–1995	74	147	221
Moved into the area before 2 years of age	6	10	16
Moved out from the area before 10 years of age	1	3	4
Died before 10 years of age	2	1	3
Not participating in the prevention programme	9	14	23
Number of children in the present study	68	139	207

Table 2 Characteristics of eight children in the control group (case 1–8) and 10 children in the study group (case 9–18) with WS

Case no.	Year of birth	CP subtype[a]	GMFCS[b]	Age at WS	Age at scoliosis	Age at hip dislocation	Age at hip osteotomy[c]
1	1990	D	3	9.5	–	–	–
2		D	4	7.5	9.0	6.0	–
3		D	4	8.5	5.0	–	–
4		D	4	6.5	6.5	–	–
5		T	5	8.5	8.5	7.0	–
6		T	5	6.0	5.5	5.0	–
7		T	5	7.0	–	5.0	–
8		T	5	7.0	7.0	8.0	–
9	1992	D	4	8.5	–	–	6.5
10		D	4	7.5	3.0	–	7.0
11		Dy	4	4.0	–	–	–
12		D	5	2.0	–	–	5.5
13		Dy	5	7.5	–	–	6.0
14		Dy	5	4.0	–	–	4.0
15		T	5	8.0	9.0	–	–
16		T	5	6.0	5.0	–	7.0
17		T	5	5.5	–	–	6.0
18		T	5	4.5	2.5	–	6.0

WS, windswept hip deformity; CP, cerebral palsy; D, spastic diplegia; T, spastic tetraplegia; Dy, dystonic type. [a]According to Hagberg *et al.* [5] [b]Gross Motor Function Classification System (GMFCS) according to Palisano *et al.* [6]. [c]Varisation osteotomy of proximal femur.

to be included as WS in this study. In the registration of scoliosis, we included those with a Cobb angle of 20° or more. From the database, the number of children operated on with varisation osteotomy of the proximal femur was noted.

Fisher's exact test was used to compare the number of children with WS, and the number of children with WS in combination with hip dislocation and scoliosis in the two age groups.

Results

WS developed in eight of 68 (12%) children in the control group. Of the eight children, six had also developed scoliosis and five had developed hip dislocation before 10 years of age (Table 2). One of the two children born during 1990–1991, excluded because of death at 9 years of age, had WS, hip dislocation and scoliosis.

Among the children in the study group, 10 out of 139 (7%) developed WS. Of the 10 children, four also developed scoliosis. These children were included in the hip prevention programme, and none developed hip dislocation. Eight of the children, however, were operated on with varisation osteotomy of the proximal femur, and in three of these cases the varisation caused a decrease in abduction, making the children fulfil the criteria for inclusion as WS. The statistical comparison between the control group and the study group is shown in Table 3.

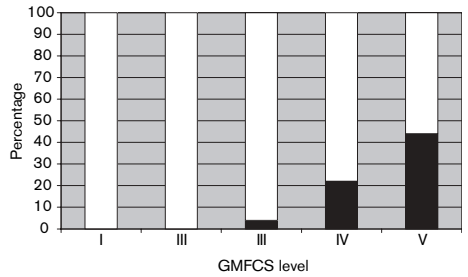
Table 3 Number of children with windswept hip deformity (WS) and with WS in combination with scoliosis (S) and hip dislocation (HD) in the control group and study group

	Control group	Study group	*P*
WS	8	10	0.20
WS + S	6	4	0.067
WS + HD	5	0	0.003
WS + S + HD	4	0	0.011

The children with WS had CP subtype spastic diplegia, spastic tetraplegia and the dystonic type. All but one were in GMFCS levels IV and V. The frequency related to the total population of children with CP is shown in Figs 1 and 2.

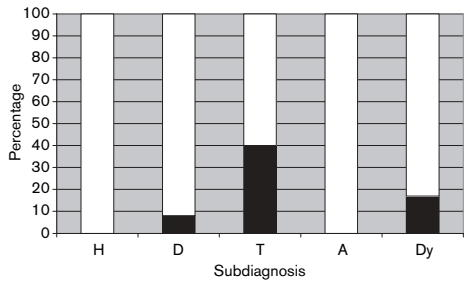
Scoliosis with Cobb angle $> 20°$ was seen before or simultaneously with WS in four out of eight children in the control group and in three out of 10 children in the study group. Hip dislocation was seen before WS in four out of eight children in the control group (Table 2).

Fig. 1



Percentage of children with windswept hip deformity (WS) in a total population of children with cerebral palsy born during 1990–1995, and in relation to Gross Motor Function Classification System (GMFCS) level. Black, WS; white, not WS.

Fig. 2



Percentage of children with windswept hip deformity (WS) in a total population of children with cerebral palsy (CP) born during 1990–1995, and in relation to CP subtype. Black, WS; white, not WS; H, spastic hemiplegia; D, spastic diplegia; T, spastic tetraplegia; A, ataxic type; Dy, dyskinetic type.

Using the Ashworth scale, seven of the 18 children with WS had a higher muscle tone in the adductor muscles on the adducted side than on the contralateral abducted side.

Discussion

The frequency of WS was 12% in the control group and 7% in the study group. The children with WS in the study group had a lower frequency of scoliosis and hip dislocation. It thus seems that the hip prevention programme results in a decrease in the number of children with WS, and a decrease in the severity of deformity.

Young *et al.* [8] constructed a formula for definition of WS. A ratio of hip medial to lateral rotation or adduction to abduction of less than 0.5 or greater than 2.0 was defined as WS. Using this formula, they found a prevalence of WS in 52% of children with the quadriplegic type of CP. As one-third of all children with CP have quadriplegic CP [8], this corresponds to a prevalence of about 17% in a total population.

Letts *et al.* [1] analysed the temporal relationship of hip dislocation, scoliosis and pelvic obliquity in the development of WS. Among children developing hip dislocation, pelvic obliquity and scoliosis, the most common sequence was hip dislocation, followed by pelvic obliquity, and finally scoliosis. This sequence was seen in three out of four cases. In the present material, using our definitions of WS and scoliosis, the WS was preceded by scoliosis in seven of 18 children. It thus seems that we have a higher proportion of children with WS starting with scoliosis. The decreasing total number of WS in the study group could be explained by the fact that the hip prevention programme has resulted in a prevention of WS starting in the hips, but not the number of WS starting with scoliosis.

Young *et al.* [8] showed a tonal asymmetry where the dislocated or adducted hip had a stronger tonus in 54% of their material. In our sample, it was seen in seven out of 18 children. It is important to treat hip and knee contractures as early as possible, also in children without walking capacity. A knee contracture combined with asymmetric tone in the adductor muscles may cause tilting of the legs to one side in lying and sitting position, and may start the development of WS.

Three children were made windswept by varisation osteotomy of the proximal femur. It seems as if this is the price one has to pay in some cases in order to save the hip from dislocation [9].

WS is very difficult to treat once the deformity is established [10,11]. Owers *et al.* [11] presented the results of treating 30 children with WS with bilateral osteotomy and soft-tissue release. They found improvement in patient handling and pain relief, but the total range of motion in the hips and the difference in abduction and adduction between the left and right hip did not improve significantly.

In conclusion, WS is a severe problem, affecting about 10% of children with CP in a total population. With a hip prevention programme, and early treatment of contracture and spasticity the frequency and severity of WS can be reduced.

References

1 Letts M, Shapiro L, Mulder K, Klassen O. The windblown hip syndrome in total body cerebral palsy. *J Pediatr Orthop* 1984; **4**:55–62.

2 Hägglund G, Andersson S, Düppe H, Lauge Pedersen H, Nordmark E, Westbom L. Prevention of severe contractures might replace multilevel surgery in cerebral palsy: results of a population based health care programme and new techniques to reduce spasticity. *J Pediatr Orthop B* 2005; **14**:268–272.
3 Hägglund G, Andersson S, Düppe H, Lauge Pedersen H, Nordmark E, Westbom L. Prevention of dislocation of the hip in children with cerebral palsy. The first ten years of a population-based programme. *J Bone Joint Surg* 2005; **87B**:95–101.
4 Nordmark E, Hägglund G, Lagergren J. Cerebral palsy in southern Sweden. I. Prevalence and clinical features. *Acta Paediatr* 2001; **90**: 1271–1276.
5 Hagberg B, Hagberg G, Olow I. The changing panorama of cerebral palsy in Sweden 1954–1970. *Acta Paediatr Scand* 1975; **64**:187–192.
6 Palisano R, Rosenbaum P, Walter S, Russel D, Wood E, Galuppi B. Development and reliability of a system to classify gross motor function in children with cerebral palsy. *Dev Med Child Neurol* 1997; **39**: 214–223.
7 Bohannon RW, Smith MB. Inter-rater reliability of a modified Ashworth scale of muscle spasticity. *Phys Ther* 1987; **67**:206–207.
8 Young NL, Wright JG, Lam P, Rajaratnam K, Stephens D, Wedge JH. Windswept hip deformity in spastic quadriplegic cerebral palsy. *Pediatr Phys Ther* 1998; **10**: 3.
9 Noonan KJ, Walker TL, Kayes KJ, Feinberg J. Varus derotation osteotomy for the treatment of hip subluxation and dislocation in cerebral palsy: statistical analysis in 73 hips. *J Pediatr Orthop B* 2001; **10**:279–286.
10 Abel MF, Blanco JS, Pavlovich L, Damiano DL. Asymmetric hip deformity and subluxation in cerebral palsy: an analysis of surgical treatment. *J Pediatr Orthop* 1999; **19**:479–485.
11 Owers KL, Pyman J, Gargan MF, Witherow PJ, Portinaro NMA. Bilateral hip surgery in severe cerebral palsy. *J Bone Joint Surg Br* 2001; **83-B**:1161–1167.

Study II

Spine
DEFORMITY
SPINE Volume 37, Number 12, pp E708–E713

Scoliosis in a Total Population of Children With Cerebral Palsy

Måns Persson-Bunke, MD,* Gunnar Hägglund, MD, PhD,* Henrik Lauge-Pedersen, MD, PhD,* Philippe Wagner, MA,† and Lena Westbom, MD, PhD‡

Study Design. Epidemiological total population study based on a prospective follow-up cerebral palsy (CP) registry.

Objective. To describe the prevalence of scoliosis in a total population of children with CP, to analyze the relation between scoliosis, gross motor function, and CP subtype, and to describe the age at diagnosis of scoliosis.

Summary of Background Data. Children with CP have an increased risk of developing scoliosis. The reported incidence varies, partly due to different definitions and study groups. Knowledge of the prevalence and characteristics of scoliosis in an unselected group of children with different CP types and levels of function is important for health care planning and for analyzing the risk in an individual child.

Methods. A total population of 666 children with CP, aged 4 to 18 years on January 1, 2008, followed with annual examinations in a health care program was analyzed. Gross Motor Function Classification System (GMFCS) level, CP subtype, age at clinical diagnosis of scoliosis, and the Cobb angle at the first radiographical examination were registered.

Results. Of the 666 children, 116 (17%) had mild and another 76 (11%) had moderate or severe scoliosis based on clinical examination. Radiographical examination showed a Cobb angle of more than 10° in 54 (8%) children and a Cobb angle of more than 20° in 45 (7%) children. The risk of developing scoliosis increased with GMFCS level and age. In most children, the scoliosis was diagnosed after 8 years of age. Children in GMFCS level IV or V had a 50% risk of having moderate or severe scoliosis by 18 years of age, whereas children in GMFCS level I or II had almost no risk.

Conclusion. The incidence of scoliosis increased with GMFCS level and age. Observed variations related to CP subtype were confounded by the GMFCS, reflecting the different distribution of GMFCS levels in the subtypes. Follow-up programs for early detection of scoliosis should be based on the child's GMFCS level and age.

Key words: cerebral palsy, scoliosis, prevalence, total population.
Spine 2012;37:E708–E713

Children with cerebral palsy (CP) have an increased risk of developing scoliosis.[1,2] The reported prevalence varies between 15% and 80% depending on different definitions of scoliosis and variations in age and severity of CP in the populations studied.[3–5] Most studies are based on children with severe impairment. There is, to our knowledge, no previous study on the prevalence of scoliosis in a total population of children with CP.

Scoliosis has been associated with problems sitting, pressure ulcers, cardiopulmonary dysfunction, gastrointestinal dysfunction, and pain.[6–11] It has also been shown to be associated with pelvic obliquity, windswept deformity, and hip dislocation.[6,12,13]

In children with CP, a spinal brace may slow the rate of progression of the curve magnitude.[14] But most curves with a Cobb angle exceeding 40° will progress, also in adulthood, if not treated surgically.[15] The outcome of spinal surgery is strongly correlated to the curve magnitude,[5] implying that early diagnosis of a scoliosis needing operation is important.

Knowledge of the prevalence and incidence of scoliosis in an unselected group of children is of interest for health care planning, for predicting future risk in a young child with CP, and for creating surveillance programs for scoliosis in children with CP.

Since 1994, there has been a follow-up health care program and registry (CPUP) for children with CP in the south of Sweden, a region with about 1.3 million inhabitants.[16,17] The prevalence of CP was 2.4 and 2.7 per 1000 children in 1998 and 2002, respectively.[18,19] Almost all (98.5%) children with CP in the area participate in the CPUP.[19] The program includes yearly spinal examinations. The purposes of this study were to describe the prevalence of scoliosis in children with CP, to analyze the relation between scoliosis incidence and the level of gross motor function and CP subtype, and to describe the age at diagnosis of scoliosis.

*Department of Orthopaedics and; †Swedish National Competence Centre for Musculoskeletal Disorders, Lund University Hospital; and ‡Division of Paediatrics, Department of Clinical Sciences, Lund University, Lund, Sweden.

Acknowledgment date: April 6, 2011. First revision date: August 19, 2011. Second revision date: December 9, 2011. Acceptance date: December 14, 2011.

The manuscript submitted does not contain information about medical device(s)/drug(s).

Institutional and foundation funds were received in support of this work.

No benefits in any form have been or will be received from a commercial party related directly or indirectly to the subject of this manuscript.

Address correspondence and reprint requests to Måns Persson-Bunke, MD, Department of Orthopaedics, Lund University Hospital, SE-221 85 Lund, Sweden; E-mail: mans.persson-bunke@skane.se

DOI: 10.1097/BRS.0b013e318246a962

TABLE 1. Distribution of Scoliosis in Relation to the GMFCS Level

GMFCS Level	Total Population	Clinical Scoliosis		Cobb Angle*			
		Mild	Mod/Severe	0°–10°	11°–20°	21°–40°	> 40°
I	306	56	5	3	0	0	0
II	86	22	5	1	2	1	0
III	80	13	7	0	1	2	0
IV	87	13	18	3	2	11	3
V	107	12	41	6	4	15	13
Total	666	116	76	13	9	29	16

**Cobb angle denotes result of first examination after diagnosis in 67 of the children with moderate or severe scoliosis.*

GMFCS indicates Gross Motor Function Classification System; Mod, moderate.

MATERIAL AND METHODS

All children with CP in the region, participating in CPUP, and born between January 1, 1990, and December 31, 2004, were included. Children who died before the age of 5 years or who had moved out of the area before the age of 5 years were excluded. Children with CP not participating in the program (1.5%) were known; no signs of dropout bias were observed. The assessments were performed from July 1, 1995, until December 31, 2008.

CPUP includes a program for monitoring scoliosis. The participating children are examined by their local physiotherapist in a standardized way twice a year from the inclusion in the program, usually at 2 years of age until 6 years of age, and then once a year. The clinical examination of the spine is done in standing position if possible; otherwise, it is performed in sitting position. The spine is examined in the extended position and with the forward bending test. The degree of scoliosis is graded as mild (discrete curve visible on thorough examination), moderate (obvious curve in both extended position and forward bending test), or severe (pronounced curve preventing upright positioning without external support). Guidelines for the clinical examination are outlined in a manual linked to the recording form. The results of the clinical measurements are registered in a database. Surgical treatment of scoliosis is also documented.

In the program, all children younger than 8 years with a nonflexible scoliosis and all children older than 8 years with a moderate or severe scoliosis are examined radiographically with anteposterior and lateral views of the entire spine. The radiographical examination is done in standing or sitting position if possible; otherwise in supine position. The localization of scoliosis and the curve magnitude measured as Cobb angle[20] are registered. Further radiographical examinations are based on the progression of the Cobb angle and the age of the child.

Gross motor function for each child was determined by the child's physiotherapist, using the Gross Motor Function Classification System (GMFCS),[21] a 5-level system for children and adolescents with CP, based on self-initiated movement, where level I describes the highest level of function and level V the lowest.

CP was defined according to Mutch *et al.*[22] CP subtype was classified according to the Surveillance of Cerebral Palsy in Europe network as spastic unilateral, spastic bilateral, dyskinetic, ataxic, or mixed type.[23]

In this study, we used the clinical and radiographical data from the registry to identify all children with scoliosis in the area during July 1, 1995, until December 31, 2008. The period prevalence was calculated, and the children with scoliosis were compared with the total population of children with CP in the registry.

Linear regression analysis was used to evaluate the effect of CP subtype and GMFCS on the Cobb angle at the first radiographical examination. GMFCS level I and spastic unilateral CP were used as reference categories. The Kaplan-Meyer analysis was used to identify the age at diagnosis of a moderate or severe scoliosis. The curves illustrate the probability of not being diagnosed with scoliosis over time in different GMFCS levels. Cox regression analysis was used to compare the incidence of scoliosis in different age groups and GMFCS levels.

The study was approved by the Medical Research Ethics Committee at Lund University (LU-433-99).

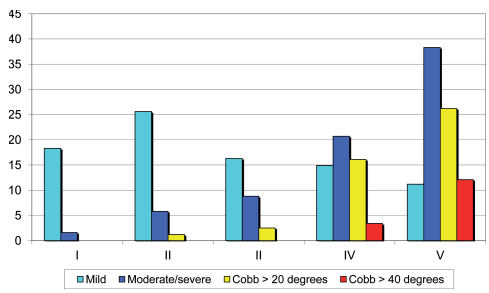


Figure 1. Scoliosis in relation to the GMFCS level. Distribution of scoliosis (%) according to clinical examination and first radiographical examination. Children with a Cobb angle of more than 40° are also included in the group with a Cobb angle of more than 20°.

TABLE 2. Distribution of Scoliosis in Relation to Cerebral Palsy Subtype

Cerebral Palsy Subtype	Total Population	Clinical Scoliosis		Cobb Angle*			
		Mild	Mod/Severe	0°–10°	11°–20°	21°–40°	> 40°
S-unilateral	192	35	4	2	1	0	0
S-bilateral	244	49	48	7	5	18	12
Ataxic	75	9	5	1	1	2	1
Dyskinetic	66	18	16	3	1	7	3
Unclassified	89	5	3	0	1	2	0
Total	666	116	76	13	9	29	16

*Cobb angle denotes the result of the first examination after diagnosis in 67 of the children with moderate or severe scoliosis.

S indicates spastic; Mod, moderate.

RESULTS

The study was based on 7200 measurements in 666 children with CP. Of these, 192 children had scoliosis according to the physiotherapists' reports, 116 were graded as mild, and 76 as moderate or severe.

Of the 76 children (50 boys and 26 girls) with moderate or severe scoliosis, 67 children were examined radiographically. The Cobb angle at the first radiographical examination after clinical diagnosis was less than 10° or less in 13 children, 11° to 20° in 9 children, 21° to 40° in 29 children, and more than 40° in 16 children (Table 1). The scoliosis was thoracolumbar in 48 children (22 left, 14 right, and 12 S-shaped), lumbar in 5 children, and thoracic in 2 children. In 9 children with moderate/severe scoliosis, no radiographical examination was performed. Two of these children died before examination, and 1 child was considered to be in too bad condition for operation, and thereby not examined radiographically. The remaining 6 children were all in GMFCS levels II–III. They were, for unknown reasons, not immediately referred for radiographical examination after having their scoliosis graded as moderate; perhaps, there was a second opinion. The further clinical examinations again graded their scoliosis as mild, and the children have therefore not been examined radiographically.

Eighteen children were operated on for scoliosis. The median age at surgery was 13 years (range, 8–17). The mean preoperative Cobb angle was 69° (median, 67; range, 40–95). All 9 children with hip dislocation, defined as migration percentage[24] of 100%, had moderate-severe scoliosis. Eight of the 9 children with hip dislocation were examined radiographically and had Cobb angles of 70° to 102°, and 4 of these children were operated on for scoliosis.

The proportion of children with scoliosis increased with GMFCS level (Table 1, Figure 1). Almost all children with moderate or severe clinical scoliosis or radiographical scoliosis with curves of more than 20° were in GMFCS levels III–V. All children operated on for scoliosis were in GMFCS levels IV–V.

The proportion of children with scoliosis varied between CP subtypes (Table 2 and Figure 2). No child with spastic unilateral CP and 3 of 75 children with ataxic CP had a curve of more than 20°. Thirty-eight of the 244 children with spastic bilateral CP (16%) and 10 of the 66 children with dyskinetic CP (15%) had a curve of more than 20°. The children operated on for scoliosis had spastic bilateral CP (15 children) and dyskinetic CP (3 children).

The linear regression estimates showed that the GMFCS level was the only statistically significant ($P = 0.004$) risk factor that affected the magnitude of Cobb angle at the first radiographical examination (Table 3).

Kaplan-Meier survival estimations showed that scoliosis was diagnosed after 8 years of age in most of the children (Figure 3). The risk of scoliosis increased with age and GMFCS level. The risk of having moderate or severe scoliosis in children in GMFCS levels IV–V was about 50% at 18 years of age. Cox regression analysis showed that a high GMFCS level indicated a high risk of scoliosis (Table 4). No significant differences were found in the analysis regarding CP subtypes.

DISCUSSION

Knowledge of the real prevalence and incidence of scoliosis in children with CP is important, both for health care planning and for analyzing the future risk for scoliosis in an individual

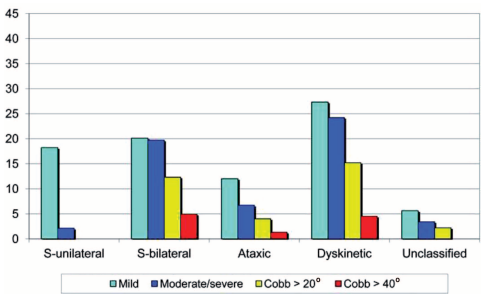


Figure 2. Scoliosis in relation to CP subtype. Distribution of scoliosis (%) according to clinical examination and first radiographical examination. Children with a Cobb angle of more than 40° are also included in the group with a Cobb angle of more than 20°. S indicates spastic.

TABLE 3. Linear Regression Estimates of the Effect of Age, GMFCS Level, and Cerebral Palsy Subtype on the Magnitude of Cobb Angle at Scoliosis Diagnosis, With 95% CI and *P* Value, in the 67 Children Radiographically Examined

	Coefficient	95% CI	*P*
GMFCS	8.09	2.78–13.39	0.004
S-bilateral	−9.17	−34.70 to 16.37	0.471
Ataxic	−12.67	−58.01 to 32.68	0.574
Dyskinetic	−21.11	−49.28 to 7.05	0.137
Age	−0.20	−2.04 to 1.65	0.830

GMFCS I and S-unilateral cerebral palsy were used as reference categories. Too few cases for analysis in children with unclassified cerebral palsy.

GMFCS indicates Gross Motor Function Classification System; CI, confidence interval; S, spastic.

child. To be able to compare prevalence figures from different studies, the selection of study group, case mix, and the definitions of scoliosis must be known. We studied the prevalence of scoliosis in a well-defined total population of children with CP, 4 to 18 years of age, which is a major strength. The nonparticipating children with CP (1.5% of the population) were known and not biasing the results.[16–19] All children were followed with repeated clinical examinations in a follow-up program, and those with moderate and severe scoliosis also with radiographical examination.

The definition of scoliosis in the study may be a limitation. In the CPUP program, the indication for radiographical examination is based on the result of the clinical examinations by physiotherapists. Some children with a diagnosis of mild scoliosis and not examined radiographically could have a curve of more than 20°. It seems, however, from the results of the radiographical examinations of those with moderate/severe scoliosis that the clinical examinations do not underestimate the degree of scoliosis.

Including all children with scoliosis according to the clinical examinations corresponds to a prevalence of scoliosis of 29%. Including only those with moderate/severe scoliosis corresponds to a prevalence of 11%. Using radiographical criteria, 8% of the children had a Cobb angle of more than 10° and 7% had a Cobb angle of more than 20°. These figures could be compared with a Swedish study of the prevalence of idiopathic scoliosis in school screening.[25] Among 17,181 children aged 7 to 16 years, 2.5% had a Cobb angle of more than 10°. More than half of all children with CP have mild gross motor function limitation (GMFCS levels I–II), and this large group thus seems to have no higher risk of developing scoliosis than children without CP. Comparison with other studies of scoliosis in CP is difficult because they predate the GMFCS and represent selected groups of children. In a radiological

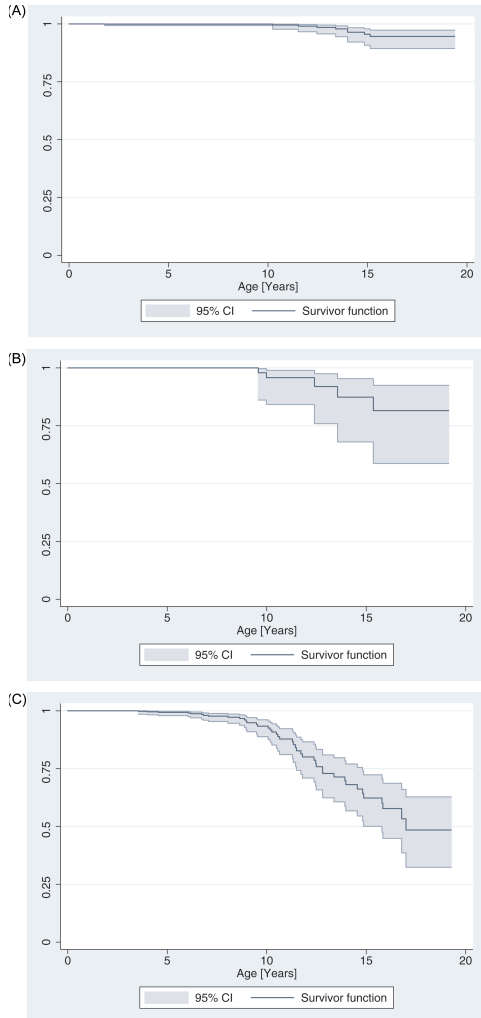


Figure 3. Survival function with 95% confidence interval (CI) illustrating the risk of having a moderate/severe scoliosis diagnosed at different ages and GMFCS levels: **(A)** GMFCS levels I–II; **(B)** GMFCS level III; and **(C)** GMFCS levels IV–V.

review of 100 children with CP from an outpatient clinic, Balmer and MacEwen[4] found 21 children with scoliosis with a curve of more than 10°. Madigan and Wallace[2] found scoliosis with a curve of more than 10° in 64% of participants from a study group of 272 institutionalized teenagers with CP. As these studies predate the GMFCS system and do not

TABLE 4. Cox Regression Analysis of the Risk Ratio (*i.e.*, Hazard Ratio) for Developing Clinically Moderate/Severe Scoliosis in Relation to the GMFCS Level and Cerebral Palsy Subtype

	Risk Ratio	95% CI	*P*
GMFCS level			
II	2.70	0.68–10.65	0.156
III	6.04	1.52–23.99	0.011
IV	14.94	4.47–49.95	<0.001
V	34.99	10.74–113.98	<0.001
S-bilateral	0.85	0.27–2.71	0.787
Ataxic	0.79	0.17–3.35	0.749
Dyskinetic	0.53	0.14–1.92	0.330
Unclassified	0.52	0.09–3.11	0.473

GMFCS I and S-unilateral cerebral palsy were used as reference categories.

GMFCS indicates Gross Motor Function Classification System; CI, confidence interval; S, spastic.

represent a total population of children with CP, the prevalence figures in the different studies are difficult to compare.

The main goals of CPUP are to prevent development of hip dislocation and severe contractures. All children at GMFCS levels III–V are examined with hip radiograph at least once a year, and children at GMFCS level II have radiographical examination at 2 and 6 years of age. In the program, the number of children with hip dislocation and windswept deformity has been reduced.[13,16] The 9 children with hip dislocation in this population had their dislocation before entering the CPUP program. In areas without hip prevention programs, the prevalence of scoliosis is presumably higher than in the present study.

The prevalence of scoliosis and the risk of being diagnosed with a moderate or severe scoliosis were related to the GMFCS level. There was also a variation in the prevalence and incidence of scoliosis with the CP subtypes. However, the Cox regression analysis revealed that the variation was explained by the different proportions of GMFCS levels in the CP subtypes used. This means that the high number of scoliosis in children with dyskinetic or spastic bilateral CP can be explained by the high number of children with GMFCS levels IV and V in those subtypes. Consequently, the low figure for scoliosis in children with spastic unilateral CP can be explained by the high number of children in GMFCS levels I–II in that subtype. This has clinical implications when analyzing the risk of scoliosis in an individual child. For this purpose, the child's GMFCS level should be used and not the CP subtype.

In most children, the scoliosis was diagnosed after 8 years of age. This is in agreement with earlier studies.[15] The highest risk of scoliosis was seen in the oldest children in GMFCS level IV or V, where about 50% were estimated to have moderate or severe scoliosis. Because there is a risk of progression of scoliosis in adulthood as well,[25] this number may increase. In the CPUP program, the children will also be followed with regular spinal examination in adulthood.

In conclusion, in this total population of children with CP, the risk of scoliosis increased with GMFCS level and age. Children in GMFCS levels I–II had almost no risk of developing scoliosis, whereas at 18 years of age, the risk among children in GMFCS levels IV–V was about 50%. Observed variations related to CP subtype were confounded by the GMFCS, reflecting the different distribution of GMFCS levels in the subtypes. Surveillance programs for scoliosis in CP should be based on the children's age and GMFCS level.

➢ Key Points

- ❑ The risk of developing scoliosis is related to the child's GMFCS level and age.
- ❑ CP subtype is not a risk factor *per se* for scoliosis.
- ❑ Children in GMFCS levels I–II have almost no risk of developing scoliosis.
- ❑ Children in GMFCS levels IV–V have a 50% risk of having clinically moderate or severe scoliosis at 18 years of age.

Acknowledgments

The study was supported by the Medical Faculty, Lund University, and the Linnéa and Josef Carlsson Foundation.

References

1. Sarwark J, Sarwahi V. New strategies and decision making in the management of neuromuscular scoliosis. *Orthop Clin North Am* 2007;3884:485–96.
2. Madigan RR, Wallace SL. Scoliosis in the institutionalized cerebral palsy population. *Spine* 1981;6:583–90.
3. Samilson RL, Bechard R. Scoliosis in cerebral palsy: incidence, distribution of curve patterns, natural history and thoughts on etiology. *Curr Pract Orthop Surg* 1973;5:183–205.
4. Balmer GA, MacEwen GD. The incidence and treatment of scoliosis in cerebral palsy. *J Bone Joint Surg Br* 1970;52B:134–7.
5. Koop SE. Scoliosis in cerebral palsy. *Dev Med Child Neurol* 2009;51(suppl 4):92–8.
6. Loeters MJ, Maathius CG, Hadders-Algra M. Risk factors for emergence and progression of scoliosis in children with cerebral palsy: a systematic review. *Dev Med Child Neurol* 2010;52:605–11.
7. Majd ME, Muldowny DS, Hort RT. Natural history of scoliosis in the institutionalized adult cerebral palsy population. *Spine* 1997;22:1461–6.
8. Yazici M, Senaran H. Cerebral palsy and spinal deformities. *Acta Orthop Traum Turc* 2009;43:149–55.
9. Koumbourlis AC. Scoliosis and the respiratory system. *Paediatr Respir Rev* 2006;7:152–60.
10. Alves VL, Avanzi O. Objective assessment of the cardiorespiratory function of adolescents with idiopathic scoliosis through the six-minute walk test. *Spine* 2009;34:E926–9.
11. Delgadillo X, Belpaire-Dethiou MC, Chantrain C, et al. Arteriomesenteric syndrome as a cause of duodenal obstruction in children with cerebral palsy. *J Pediatr Surg* 1997;32:1721–3.

12. Lonstein JE. The spine in cerebral palsy. Mini-symposium: cerebral palsy. *Cur Orthop*. 1995;9:164–77.
13. Persson-Bunke M, Hägglund G, Lauge-Pedersen H. Windswept hip deformity in children with cerebral palsy. *J Pediatr Orthop B* 2006;15:335–8.
14. Terjesen T, Lange JE, Steen H. Treatment of scoliosis with spinal bracing in quadriplegic cerebral palsy. *Dev Med Child Neurol* 2000;42:448–54.
15. Saito N, Ebara S, Ohotsuka K, et al. Natural history of scoliosis in spastic cerebral palsy. *Lancet* 1998;351:1687–92.
16. Hägglund G, Andersson S, Düppe H, et al. Prevention of hip dislocation in children with cerebral palsy. The first ten years experience of a population-based prevention programme. *J Bone Joint Surg Br* 2005;87:95–101.
17. Hägglund G, Andersson S, Düppe H, et al. Prevention of severe contractures might replace multi-level surgery in CP. Results of a population based health care program and new techniques to reduce spasticity. *J Pediatr Orthop* 2005;14:268–72.
18. Nordmark E, Hägglund G, Lagergren J. Cerebral palsy in southern Sweden. Prevalence and clinical features. *Acta Paediatr* 2001;90:1271–6.
19. Westbom L, Hägglund G, Nordmark E. Cerebral palsy in a total population of 4–11 year olds in southern Sweden. Prevalence and distribution according to different CP classification systems. *BMC Pediatr* 2007:7:41.
20. Cobb JR. Outline for the study of scoliosis. In: *Instructional Course Lectures*. Vol 5. Rosemont, IL: American Academy of Orthopaedic Surgeons; 1948:261–75.
21. Palisano R, Rosenbaum P, Walter S, et al. Development and reliability of a system to classify gross motor function in children with cerebral palsy. *Dev Med Child Neurol* 1997;39:214–23.
22. Mutch L, Alberman E, Hagberg B, et al. Cerebral palsy epidemiology: where are we now and where are we going? *Dev Med Child Neurol* 1992;34:547–51.
23. Surveillance of Cerebral Palsy in Europe. Surveillance of Cerebral Palsy in Europe (SCPE): a collaboration of cerebral palsy surveys and registers. *Dev Med Child Neurol* 2000;42:816–24.
24. Thometz JG, Simon S. Progression of scoliosis after skeletal maturity in institutionalized adults who have cerebral palsy. *J Bone Joint Surg Am* 1988;70:1290–6.
25. Willner S, Udén A. A prospective prevalence study of scoliosis in southern Sweden. *Acta Orthop Scand* 1982;53:233–7.

Study III

Validity and interrater reliability of spinal assessment methods to screen for scoliosis in children and adolescents with cerebral palsy

Måns Persson-Bunke[1], Tomasz Czuba[2], Gunnar Hägglund[1], Elisabet Rodby-Bousquet[1,3]

[1] Lund University, Department of Clinical Sciences, Lund, Orthopaedics , Lund, Sweden
[2] National Competence Center for Quality Registers, Lund University, University Hospital, Lund, Sweden
[3] Centre for Clinical Research, Uppsala University, County Hospital, Västerås, Sweden

ABSTRACT

Study design
Psychometric evaluation.

Objective
To evaluate the interrater reliability of the clinical spinal assessment method used in the Swedish follow-up program for cerebral palsy (CPUP) and scoliometer measurement in children with cerebral palsy (CP) and to evaluate their validity compared to radiographic examination.

Summary of background data
CPUP includes clinical examinations of the spine. The reliability and validity of the assessment method have not been studied.

Methods
Twenty-eight children (6-16 years) with CP in Gross Motor Function Classification System levels II-V were included. Clinical spinal examinations and scoliometer measurements in sitting position were performed by three independent examiners. The results were compared to the Cobb angle as determined by radiographic measurement. Interrater reliability was calculated using weighted Kappa. Concurrent validity was analyzed using the Cobb angle as gold standard. Sensitivity, specificity, area under receiver operating characteristic curves (AUC) and likelihood ratios (LR) were calculated. Cut-off values for scoliosis were set to ≥ 20° Cobb angle and ≥ 7° scoliometer angle.

Results
There was an excellent interrater reliability for both clinical examination (weighted kappa = 0.96) and scoliometer measurement (weighted kappa = 0.86). The clinical examination showed a sensitivity of 75.0% (95% CI: 19.4-99.4%), specificity of 99.8% (95% CI: 78.9-99.9%) and an AUC of 0.85 (95% CI: 0.61-1.00). The positive LR was 18.0 and the negative LR was 0.3. The scoliometer measurement showed a sensitivity of 50.0% (95% CI: 6.8-93.2%), specificity of 91.7% (95% CI: 73.0-99.9%) and AUC of 0.71 (95% CI: 0.42-0.99). The positive LR was 6.0 and the negative LR was 0.5.

Conclusions
The psychometric evaluation of the clinical assessment showed an excellent interrater reliability and a high concurrent validity compared to the Cobb angle. Clinical spinal examinations seem appropriate as a screening tool to identify scoliosis in children with CP.

Keywords
Cerebral palsy, scoliosis, reliability, validity, assessment, screening, measurement, neuromuscular scoliosis, psychometric evaluation.

INTRODUCTION

Children and adolescents with cerebral palsy (CP) have an increased risk of scoliosis (1). The reported prevalence varies between 15-64% based on age, severity of CP, and different definitions of scoliosis (1-3). This stands in contrast to idiopathic scoliosis in adolescents where the prevalence has been reported at 2-4% (4). In children with CP the risk of developing scoliosis is related to the child's gross motor function and age (1, 5). Severe scoliosis is associated with pain, sitting problems, hip dislocation, windswept deformity (1, 6), all of which may impair physical function and quality of life.

It is important to identify a progressive scoliosis early-on as the result of spinal surgery is related to the curve magnitude (7). It is desirable to have an examination tool with high sensitivity but it is also important to have high specificity to avoid unnecessary radiographic examinations.

In 1994, a follow-up program and registry for children and adolescents with CP (CPUP) was initiated in the south of Sweden, an area of approximately 1.3 million inhabitants. CPUP has been classified as a Swedish National Health Care Quality Registry since 2005 and the program is also used in Norway, Denmark, Iceland, Scotland and New South Wales, Australia. These days>95% of all children with CP in Sweden participate in CPUP (8) and it is currently expanding to include also adults with CP.

The main purpose of CPUP is to prevent hip dislocation, contractures, scoliosis and windswept deformities in individuals with CP (8-11). The program includes spinal examinations where the children undergo a standardized examination by their local physiotherapist twice a year until six years of age and then once a year. The spine is examined in forward bending and upright position with the child sitting on a plinth. In the event of scoliosis, it is graded as "mild", "moderate" or "severe". A child with a moderate or severe scoliosis is referred to radiographic examination. If the Cobb angle exceeds 40° operative treatment is considered.

A commonly used evaluation test to screen for adolescent idiopathic scoliosis is the forward bending test that measures asymmetrical rib prominence (12, 13). It has been argued that this test does not have a quantitative documentation and the efficacy of the test to screen for scoliosis is still discussed (12, 13).

The scoliometer (14) reliably measures the angle of trunk rotation and it is a commonly used tool to screen for adolescent idiopathic scoliosis (13, 15). The recommended cut-off value to warrant a radiographic referral in adolescent idiopathic scoliosis varies but ≥7° has been suggested (15). To our knowledge the scoliometer has not been used in screening for neuromuscular scoliosis.

The Cobb angle has been the gold standard to quantify scoliosis on radiographic examination since 1948 (16). However, the Cobb angle is a two-dimensional analysis of a lateral deviation of the spine while the forward bending test and the scoliometer reflect a lateral deviation and rotation of a three dimensional deformity. Coehlo et al found a correlation that was considered good ($r>0.7$, $p<0.05$) between scoliometer measurement and the Cobb angle in screening for adolescent idiopathic scoliosis (17).

For a screening tool to be useful in clinical practice the sensitivity and specificity is vital. When screening for scoliosis in CPUP the purpose is to identify all individuals requiring further radiographic examination and rule out those who does not in order to minimize the dose of radiation exposure.

Purpose

The purposes of this study were to evaluate the interrater reliability of clinical examination and scoliometer measurement, and to evaluate their sensitivity, specificity and concurrent validity as screening tools by using radiographic examination with the Cobb angle as reference.

MATERIALS AND METHODS

In CPUP, all participating children have their CP diagnosis verified by a neuropaediatrician at four years of age. CP is defined as a non-progressive brain injury which has developed before the age of two years. Motor impairment and specific neurological signs are defined and classified according to the inclusion criteria of the Surveillance of Cerebral palsy in Europe (SCPE) network (18, 19). Gross motor function is determined by the child's physiotherapist according to the expanded and revised version of the Gross Motor Function Classification System (GMFCS) (20, 21). This is a 5-level system for children and adolescents with CP that is based on their self-initiated movement where level I delineates the highest level of function and level V the lowest.

Children and adolescents ages 6-16 years at GMFCS levels II-V and enrolled in CPUP were recruited from five child rehabilitation units in southern Sweden. The participants and their families were informed about the study by their local physiotherapists and provided with invitation letters with information about the study. Written consent was obtained from all participants. Children were recruited consecutively until at least six children at each relevant GMFCS level had accepted. The decision to include six persons in each GMFCS level (except level I) was based on an earlier study evaluating the Posture and Postural Ability Scale in adults with CP (22, 23). In addition, a reliability study of the scoliometer by Bonagamba and colleagues included 24 participants and that study had enough power to satisfactorily assess reliability (24). Children at GMFCS level I, which constitutes about 40% of all children with CP, do not have a higher risk of scoliosis compared to the risk of developing idiopathic scoliosis in adolescents (1) and were therefore not included in this study.

The children were examined at one occasion by three examiners, independent of each other, during a period from November 2013 to March 2014. The examinations were performed by two physiotherapists and one paediatric orthopaedic surgeon, all with several years experience working with children with CP. The spine was examined with the child in a sitting upright position, with external support if needed, and then, still in sitting, with the forward bending. The degree of scoliosis was noted according to the CPUP classification (1) and graded as:

-*No* scoliosis.
-*Mild scoliosis*: discreet curve visible only on thorough examination in forward bending.
-*Moderate scoliosis*: obvious curve in both extended and forward bending.
-*Severe scoliosis*: pronounced curve preventing upright position without external support.

In sitting position, a scoliometer was placed in forward bending at the top of the thoracic spine, with the 0 (zero) mark over the spinous process, and slowly moved down the spine noting the highest degree

of truncal rotation. The degree of scoliosis was recorded separately and independently by the three examiners. A higher degree of truncal rotation indicates worse inclination. The value used to detect moderate scoliosis that should be referred to radiographic examination was set to ≥7°.

Radiographic examinations were performed in a sitting position, in an anteposterior (AP) projection. The magnitude of the curve was determined based on the Cobb angle (16) and moderate or severe scoliosis was defined as Cobb angle ≥20°.
Ethical approval was granted by the Medical Research Ethics Committee at Lund University, Dnr 467/2013.

Statistical analysis
Interrater reliability for the clinical spinal examination and the scoliometer measurement was calculated using weighted Kappa scores (25). The magnitude of weighted Kappa was interpreted according to Fleiss 1981 where ≤ 0.40 signifies poor agreement, 0.40-0.75 fair to good agreement and ≥ 0.75 signifies excellent agreement (26). To calculate 95% CI for weighted kappa scores all GMFCS levels included were combined and 95% nonparametric bootstrap confidence intervals were added based on a 1000 re-samples (27, 28).

To evaluate concurrent validity the Cobb angle was used as gold standard. Area under receiver operating characteristic curves (AUC), sensitivity, specificity and predictive values were calculated. The cutoff point chosen for clinical assessment was no or mild scoliosis versus moderate or severe scoliosis. We used averaged ratings for analyzing validity of the scoliometer but not for calculation of Kappa values.
The AUC is a measure of the capacity of a test to classify a person correctly. In this study the AUC was used as a measure of the capacity to correctly identify scoliosis according to our definition. A value of <0.5 is not better than random, > 0.7 is acceptable, >0.8 is excellent, and >0.9 is extraordinary capability (29).

Likelihood ratio (LR) is a summary of the diagnostic accuracy of a test telling the ratio of the probability of a certain test result in individuals who do have the disease to the probability in individuals who do not. The definition of a positive LR is sensitivity/ 1-specificity. The definition of a negative LR is 1-sensitivity/ specificity. A positive LR greater than 10 means that the test is good at confirming scoliosis. A negative LR less than 0.1-0.2 means that the test is good at ruling out scoliosis (30). For all statistical computing R software environment version 3.0.0 and STATA version 13.1 were used.

RESULTS

In total, 28 children with CP (14 boys), with a median age of 12 years (range 6-16 years) were included. All 28 children completed both the clinical and radiographic examinations. There were nine children in GMFCS II, seven in GMFCS III, six in GMFCS IV and six in GMFCS V (Table 1).

Clinical assessment
In 25 of the 28 children there was a total interrater agreement. Among these 25 children 14 had no scoliosis, 7 mild, 2 moderate, and 2 had severe scoliosis (Table 1). The 21 children with no or mild scoliosis had an average Cobb angle of 11° (range 0-21°). The 4 children with moderate or severe scoliosis had an average Cobb angle of 27° (range 9-38°). Interrater disagreement occurred in 3 cases (cases 9, 16 and 28; Table 1).

Table 1. Details of all participants and distribution of scores for the 3 raters (A, B, C) versus Cobb-angle.

Case Number	Age	GMFCS	Sex	Clinical examination			Scoliometer			Cobb-angle
				A	B	C	A	B	C	
1	6	IV	F	No	No	No	1	1	3	16
2	7	III	M	No	No	No	1	3	3	9
3	7	V	F	No	No	No	2	2	3	13
4	9	III	M	No	No	No	1	0	1	13
5	9	V	M	Mild	Mild	Mild	5	5	3	12
6	10	III	F	No	No	No	4	2	1	16
7	10	V	M	No	No	No	3	2	2	8
8	10	II	M	No	No	No	1	1	2	10
9	10	III	M	Mild	No	Mild	4	3	6	7
10	10	II	F	No	No	No	2	2	3	0
11	10	III	F	Moderate	Moderate	Moderate	5	6	5	23
12	10	V	M	Severe	Severe	Severe	13	18	12	37
13	11	IV	F	No	No	No	2	0	3	14
14	12	II	F	Mild	Mild	Mild	5	7	5	13
15	12	V	F	Mild	Mild	Mild	5	2	6	21
16	12	II	M	Mild	No	No	5	1	1	11
17	12	II	M	No	No	No	1	1	2	0
18	12	II	F	Mild	Mild	Mild	2	3	3	13
19	13	IV	M	Mild	Mild	Mild	4	5	5	15
20	13	II	F	No	No	No	2	2	3	13
21	13	IV	M	Severe	Severe	Severe	14	20	15	38
22	14	V	F	Mild	Mild	Mild	5	8	7	7
23	15	II	M	Mild	Mild	Mild	5	5	5	3
24	15	III	M	No	No	No	1	2	1	17
25	16	IV	F	No	No	No	1	2	2	0
26	16	IV	M	No	No	No	1	1	1	19
27	16	III	F	Moderate	Moderate	Moderate	7	10	10	9
28	16	II	F	No	Mild	No	6	6	6	11

The spines were graded as no or mild scoliosis by all 3 raters. The average Cobb angle for the 21 children was 10° (range 7-11°). The interrater reliability of the clinical assessment showed a weighted Kappa value of 0.96 (95% CI: 0.82-1.00). The sensitivity was 75.0% (95% CI: 19.4%-99.4%), the specificity was 99.8% (95% CI: 78.9%-99.9%) and the AUC was 0.85 (95% CI: 0.61-1.00). The positive predictive value was 75.0% (95% CI: 19.4-99.4%) and the negative predictive value was 99.8% (95% CI: 78.9%-99.9%). The positive LR was 18.0 and the negative LR was 0.2 (Table 2-3).

Table 2. Number of positive and negative cases of clinical spinal assessment and scoliometer measurement versus radiographic Cobb angle**.** (Average ratings of 3 examiners)

		Clinical assessment			Scoliometer		
		Positive	Negative	Total	Positive	Negative	Total
Cobb angle	Scoliosis $\geq$ 20°	3	1	4	2	2	4
	No scoliosis $\leq$ 20°	1	23	24	2	22	24
	Total	4	24	28	4	24	28

Table 3. Concurrent validity of clinical spinal assessment and scoliometer measurement versus radiographic Cobb angle.

	Clinical assessment vs Cobb			Scoliometer vs Cobb		
Prevalence		95% CI			95% CI	
Sensitivity (%)	75.0	19.4	99.4	50.0	6.8	93.2
Specificity (%)	95.8	78.9	99.9	91.7	73.0	99.9
Area under curve (AUC)	0.85	0.61	1.00	0.71	0.42	0.99
Likelihood ratio (+)	18.0	2.4	133.0	6.0	1.1	31.2
Likelihood ratio (-)	0.3	0.1	1.4	0.5	0.2	1.5
Positive predictive value (%)	75.0	19.4	99.4	50.0	6.8	93.2
Negative predictive value (%)	95.8	78.9	99.9	91.7	73.0	99.9

Scoliometer

In 23 of the children all 3 examiners measured the scoliometer angle <7°. The average scoliometer angle in these children was 3° (range 0-6°) and the average Cobb angle was 12° (range 0-23°). In 3 of the children all examiners measured the scoliometer angle above the cutoff of ≥7°. measured 5° while the other two examiners measured 7° and 8° respectively. The Cobb angle was 7°. The interrater reliability of the scoliometer measurement showed a weighted Kappa value of 0.86 (95% CI: 0.64-0.92). The sensitivity was 50% (95% CI: 6.8%-93.2%), the specificity was 91.7% (95% CI: 73.0-99.9%), and the AUC was 0.71 (95% CI: 0.42-0.99)(Table 3). The positive predictive value was 50.0% (95% CI: 6.8-93.2%) and the negative predictive value was 91.7% (95% CI: 73.0%-99.9%). The positive LR was 6.0 and the negative LR was 0.5. (Table 2-3).

DISCUSSION

We found both the clinical assessment method and the scoliometer measurement to have high interrater reliability. The clinical assessment showed a higher specificity, sensitivity and a larger area under the curve compared to the scoliometer method. For both the clinical examination and the scoliometer, the negative and positive predictive values were the same as for specificity and sensitivity. The reason for this was that the outcome table was symmetrical on the diagonal with the same number of false positives and false negatives (Table 2). The predictive value could otherwise have been negatively influenced by the low prevalence of scoliosis. The informative value and the usefulness of the test were refined for the clinical assessment by analyzing the LR.

The average scoliometer angle for these 3 children was 13° (range 7-20°, Table 1). Two children were measured both below and above the cutoff. In one child (Case 14; Table 1) 2 of the examiners measured 5° and the third examiner measured 7°. The Cobb angle was 13°. In the second child (Case 22; Table 1), one examiner

A limitation of this study was the small number of children with moderate or severe scoliosis. This could explain the wide CI for sensitivity for both assessment methods. The purpose was to evaluate the screening method used to identify scoliosis in a population of children with CP and to select those in need of further investigation. The specificity of the clinical assessment was high (99.8 %) thereby reducing unnecessary referrals for radiographic examination.

There was no disagreement among the assessors regarding the rating of moderate or severe scoliosis. The higher kappa values for the clinical assessment may partially be explained by the narrow range of the scale (0-3).

The cutoff point for the scoliometer measure was set to 7°. When screening, a balance has to be struck in terms of not referring too many or too few for a radiographic examination and this is related to the sensitivity and the specificity of the test. Bunnell (15) studied the outcome of spinal screening with scoliometer in adolescent idiopathic scoliosis and recommended 7° as an appropriate referral criterion. To use 7° as the referral criterion reduced the prognostic referral rate from 12% to 3% compared to if 5° would have been the criterion for referral. When creating guidelines for assessment of adolescent idiopathic scoliosis Coelho (17) et al concluded that if a cutoff point of 5° for scoliometer measurement was used the sensitivity would be approximately100% and the specificity approximately 47%. However, if 7° was used as the cut-off instead, the sensitivity dropped to 83% while the specificity rose to 86%. The results from Coelho and colleagues informed the choice of cut-off point in this study. The purpose of screening for adolescent idiopathic scoliosis is to detect scoliosis in time to start bracing, and thereby reducing the need for surgery. In neuromuscular scoliosis, an additional purpose of screening is to find a progressive scoliosis in time for surgery. It is also important to identify postural asymmetries that induce a worse sitting position with poor head control, uneven weight distribution, and pelvic obliquity which could increase the development of contractures, windswept position, and hip dislocation (22).

The clinical assessment and the scoliometer measurement proved somewhat difficult when examining children who had problems bending forward in sitting because of decreased flexibility of spinal muscles, short hamstrings or limited hip flexion. In one person a baclofen pump prevented the child from bending forward properly. For these individuals a moderate or severe scoliosis is often apparent when sitting in an upright position. As a consequence, when the child is unable to bend forward, the examiner is only likely to miss a mild scoliosis.

Summary/Conclusion

In summary, this study showed an excellent interrater reliability for both the clinical spinal assessment used in CPUP and for the scoliometer measurements. The sensitivity was higher for the clinical assessment compared to the scoliometer measurement, while the specificity was almost the same for both methods. There was a high validity correlated to the Cobb angle measurement. The clinical spinal assessment method used in CPUP seems to be an appropriate screening method for scoliosis in children with CP.

List of abbreviations

AUC = Area under curve
CP = Cerebral palsy
CPUP = Cerebral palsy follow up programme and quality registry
GMFCS = Gross motor function classification system
LR = Likelihood ratio
PPAS = Posture and postural ability scale
SCPE = Surveillance of cerebral palsy in Europe network

Conflicts of interests

The authors declare that they have no conflicts of interests.

Acknowledgements

The study was supported by the Medical Faculty, Lund University and Stiftelsen för bistånd åt rörelsehindrade i Skåne. We would also like to thank physiotherapist Annette Sällvik for assessing all children.

Authors' contributions

MPB designed the study, performed the examinations, collected and analyzed data and drafted the manuscript. TC participated in its design, performed the

statistical analyses and actively improved and revised the manuscript. GH participated in the design, analysis of data and actively revised and improved the manuscript. ERB participated in the design, the clinical examinations, analyzes of data and actively revised and improved the manuscript. All authors approved the final draft for publication.

REFERENCES

1. Persson-Bunke M, Hagglund G, Lauge-Pedersen H, Wagner P, Westbom L. Scoliosis in a total population of children with cerebral palsy. Spine. 2012;37(12):E708-13.
2. Koop SE. Scoliosis in cerebral palsy. Developmental medicine and child neurology. 2009;51 Suppl 4:92-8.
3. Madigan RR, Wallace SL. Scoliosis in the institutionalized cerebral palsy population. Spine. 1981;6(6):583-90.
4. Ersberg A, Gerdhem P. Pre- and postoperative quality of life in patients treated for scoliosis. Acta orthopaedica. 2013;84(6):537-43.
5. Saito N, Ebara S, Ohotsuka K, Kumeta H, Takaoka K. Natural history of scoliosis in spastic cerebral palsy. Lancet. 1998;351(9117):1687-92.
6. Sarwark J, Sarwahi V. New strategies and decision making in the management of neuromuscular scoliosis. The Orthopedic clinics of North America. 2007;38(4):485-96, v.
7. Miyanji F, Slobogean GP, Samdani AF, Betz RR, Reilly CW, Slobogean BL, et al. Is larger scoliosis curve magnitude associated with increased perioperative health-care resource utilization?: a multicenter analysis of 325 adolescent idiopathic scoliosis curves. The Journal of bone and joint surgery American volume. 2012;94(9):809-13.
8. Westbom L, Hägglund G, Nordmark E. Cerebral palsy in a total population of 4-11 year olds in southern Sweden. Prevalence and distribution according to different CP classification systems. BMC pediatrics. 2007;7:41.
9. Hägglund G, Andersson S, Duppe H, Lauge-Pedersen H, Nordmark E, Westbom L. Prevention of severe contractures might replace multilevel surgery in cerebral palsy: results of a population-based health care programme and new techniques to reduce spasticity. Journal of pediatric orthopedics Part B. 2005;14(4):269-73.
10. Hägglund G, Andersson S, Duppe H, Lauge-Pedersen H, Nordmark E, Westbom L. Prevention of dislocation of the hip in children with cerebral palsy. The first ten years of a population-based prevention programme. J Bone Joint Surg Br. 2005;87(1):95-101.
11. Hägglund G, Alriksson-Schmidt A, Lauge-Pedersen H, Rodby-Bousquet E, Wagner P, Westbom L. Prevention of dislocation of the hip in children with cerebral palsy: 20-year results of a population-based prevention programme. The bone & joint journal. 2014;96-b(11):1546-52.
12. Cote P, Kreitz BG, Cassidy JD, Dzus AK, Martel J. A study of the diagnostic accuracy and reliability of the Scoliometer and Adam's forward bend test. Spine. 1998;23(7):796-802; discussion 3.
13. Amendt LE, Ause-Ellias KL, Eybers JL, Wadsworth CT, Nielsen DH, Weinstein SL. Validity and reliability testing of the Scoliometer. Physical therapy. 1990;70(2):108-17.
14. Bunnell WP. An objective criterion for scoliosis screening. The Journal of bone and joint surgery American volume. 1984;66(9):1381-7.
15. Bunnell WP. Outcome of spinal screening. Spine. 1993;18(12):1572-80.
16. Cobb J. Outline for the study of scoliosis. In Instructional Course

Lectures Rosemont I, editor: American Academy of Orthopaedic Surgeons 1948.

17. Coelho DM, Bonagamba GH, Oliveira AS. Scoliometer measurements of patients with idiopathic scoliosis. Brazilian journal of physical therapy. 2013;17(2):179-84.
18. Rosenbaum P, Paneth N, Leviton A, Goldstein M, Bax M, Damiano D, et al. A report: the definition and classification of cerebral palsy April 2006. Developmental medicine and child neurology Supplement. 2007;109:8-14.
19. Surveillance of cerebral palsy in Europe: a collaboration of cerebral palsy surveys and registers. Surveillance of Cerebral Palsy in Europe (SCPE). Developmental medicine and child neurology. 2000;42(12):816-24.
20. Palisano R, Rosenbaum P, Walter S, Russell D, Wood E, Galuppi B. Development and reliability of a system to classify gross motor function in children with cerebral palsy. Developmental medicine and child neurology. 1997;39(4):214-23.
21. Palisano RJ, Rosenbaum P, Bartlett D, Livingston MH. Content validity of the expanded and revised Gross Motor Function Classification System. Developmental medicine and child neurology. 2008;50(10):744-50.
22. Rodby-Bousquet E, Czuba T, Hägglund G, Westbom L. Postural asymmetries in young adults with cerebral palsy. Developmental medicine and child neurology. 2013;55(11):1009-15.
23. Rodby-Bousquet E, Agustsson A, Jonsdottir G, Czuba T, Johansson AC, Hagglund G. Interrater reliability and construct validity of the Posture and Postural Ability Scale in adults with cerebral palsy in supine, prone, sitting and standing positions. Clinical rehabilitation. 2014;28(1):82-90.
24. Bonagamba GH, Coelho DM, Oliveira AS. Inter and intra-rater reliability of the scoliometer. Revista brasileira de fisioterapia (Sao Carlos (Sao Paulo, Brazil)). 2010;14(5):432-8.
25. Conger AJ. Integration and generalization of kappas for multiple raters. Psychological Bulletin. 1980;88(2):322-8.
26. Fleiss J, L. , editor. Statistical Methods for Rates and Proportions. Second ed1981.
27. DiCiccio TJ, Efron B. Bootstrap Confidence Intervals. Statistical Science. 1996;11(3):189-212.
28. Davison ACaH, D.V., editor. Bootstrap Methods and Their Application, Chapter 5. : Cambridge University Press; 1997.
29. Pepe MS. The statistical evaluation of medical tests for classification and prediction. Oxford ; New York: Oxford University Press; 2003. xvi, 302 p. p.
30. Grimes DA, Schulz KF. Refining clinical diagnosis with likelihood ratios. Lancet. 2005;365(9469):1500-5.

Study IV

Article

CLINICAL REHABILITATION

Clinical Rehabilitation
1–8

DOI: 10.1177/0269215515593612
cre.sagepub.com
SAGE

Psychometric evaluation of the Posture and Postural Ability Scale for children with cerebral palsy

Elisabet Rodby-Bousquet[1,2], Måns Persson-Bunke[2] and Tomasz Czuba[3]

Abstract

Objective: To evaluate construct validity, internal consistency and inter-rater reliability of the Posture and Postural Ability Scale for children with cerebral palsy.

Design: Evaluation of psychometric properties.

Setting: Five child rehabilitation centres in the south of Sweden, in November 2013 to March 2014.

Subjects: A total of 29 children with cerebral palsy (15 boys, 14 girls), 6–16 years old, classified at Gross Motor Function Classification System (GMFCS) levels II ($n = 10$), III ($n = 7$), IV ($n = 6$) and V ($n = 6$).

Main measures: Three independent raters (two physiotherapists and one orthopaedic surgeon) assessed posture and postural ability of all children in supine, prone, sitting and standing positions, according to the Posture and Postural Ability Scale. Construct validity was evaluated based on averaged values for the raters relative to known-groups in terms of GMFCS levels. Internal consistency was analysed with Cronbach's alpha and corrected Item–Total correlation. Inter-rater reliability was calculated using weighted kappa scores.

Results: The Posture and Postural Ability Scale showed construct validity and median values differed between GMFCS levels ($p < 0.01$). There was a good internal consistency (alpha = 0.95–0.96; item–total correlation = 0.55–0.91), and an excellent inter-rater reliability (kappa score = 0.77–0.99).

Conclusion: The Posture and Postural Ability Scale shows high psychometric properties for children with cerebral palsy, as previously seen when evaluated for adults. It enables detection of postural deficits and asymmetries indicating potential need for support and where it needs to be applied.

Keywords

Posture and Postural Ability Scale, posture, postural control, cerebral palsy, reliability, validity, psychometric evaluation

Received: 21 March 2015; accepted: 7 June 2015

[1]Centre for Clinical Research, Uppsala University, County Hospital, Västerås, Sweden
[2]Department of Clinical Sciences, Lund University, Lund, Sweden
[3]National Musculoskeletal Competence Centre for Quality Registers, Lund University, Lund, Sweden

Corresponding author:
Elisabet Rodby-Bousquet, Centre for Clinical Research, Uppsala University, County Hospital, SE-721 89 Västerås, Sweden.
Email: elisabet.rodby_bousquet@med.lu.se

Introduction

Asymmetric postures can cause contractures, bone and joint deformities in immobile children with cerebral palsy.[1–4] Most of these deformities can be prevented via active surveillance and therefore identification of asymmetries and postural deficits should be used to screen for contractures.[5–9]

The Posture and Postural Ability Scale (PPAS)[10] is the only clinical assessment tool designed to assess 'quality' and 'quantity' of posture separately, in the four basic body positions: supine, prone, sitting and standing. 'Quality' of posture, relates to the shape of the body, that is, the particular alignment of body segments in relation to each other and to the supporting surface. 'Quantity' refers to postural ability, that is, the ability to stabilize the body segments relative to each other and to the supporting surface. This means control of the centre of gravity relative to the base of support during both static and dynamic conditions.[11,12] The levels of postural ability are based on the original work by Noreen Hare[13] to assess children and adolescents with severe motor impairments and scoliosis. Her Physical ability scale has been evaluated for inter-rater and intra-rater reliability in children.[13] The levels are also based on the related Chailey levels of abilities[14] evaluated for validity in children with cerebral palsy.[15] Pauline Pope modified these scales and added items for quality of posture for use with people with disabilities regardless of age and diagnosis. All three scales have been used by trained therapists in England since the 1990s. In 2011, Pope and colleagues in Iceland and Sweden expanded and revised the assessment tool into the PPAS. It has shown excellent inter-rater reliability (kappa 0.85–0.99), high internal consistency (alpha 0.96–0.97) and construct validity ($p < 0.02$) for adults with cerebral palsy when used by trained professionals, but it has not previously been evaluated for use with children or for less experienced raters. It is currently used in National follow-up programmes for adults with cerebral palsy in Sweden and Iceland.

The purpose of this study was to evaluate construct validity, internal consistency and inter-rater reliability of the PPAS for children with cerebral palsy.

Methods

Children between 6 and 16 years old who participated in the Swedish national cerebral palsy healthcare programme called CPUP,[5,16] were recruited from five child rehabilitation units in southern Sweden. Invitation letters and written information about the study was given to the families by their local physiotherapists. Written consent from all families who agreed to participate was sent to the Department of Orthopaedics at Lund University. Those who accepted were examined once during a period from November 2013 to March 2014. All children who participated had cerebral palsy verified by a neuropaediatrician, with a non-progressive brain injury before the age of 2 years, and motor impairment and specific neurological signs, defined by the inclusion criteria of the Surveillance of Cerebral Palsy in Europe (SCPE) network.[17]

Children were invited consecutively until at least six children at each level II–V of the Gross Motor Function Classification System (GMFCS)[18] had accepted. The classification has five levels based on self-initiated movement. The level of gross motor function was classified by each child's local physiotherapist. The selection of six subjects at each GMFCS level was based on a previous psychometric evaluation of the PPAS for use with adults.[10] In order to evaluate construct validity, we used known groups based on the GMFCS levels, assuming that posture is likely to be more asymmetric and postural ability more impaired in children at lower levels of motor function, such as GMFCS level IV and V. The study was approved by the Medical Research Ethics Committee at Lund University, number D467/2013.

The PPAS[10] is designed to assess postural control and asymmetries in people with severe disabilities in four basic body positions; supine and prone lying, sitting and standing. Quality of posture is rated for position of head, trunk, pelvis, legs, arms and weight distribution in the frontal plane, and the sagittal plane. Symmetry and alignment scores 1 point for each item, while asymmetry or deviation from midline scores 0 points. The total score varying from 0–6 points for each position in the frontal and the sagittal plane is calculated separately. Quantity is rated on an ordinal scale, where postural ability ranges from

'unplaceable in an aligned posture' (level 1), to 'placeable in an aligned posture but needs support' (level 2), 'able to maintain position when placed but cannot move' (level 3), 'able to initiate flexion/extension of trunk' (level 4), 'able to transfer weight laterally and regain posture' (level 5), 'able to move out of position' (level 6) and the highest level of ability 'able to move into and out of position' (level 7). It is important to note that levels 1 and 2 relate to the person with little or no postural ability. Thus it is possible to have a person with a high level of ability, that is, 'able to move into and out of position' who scores 0 for quality of posture owing to contracture, deformity or strategies used to gain stability.

All children were examined at their local child rehabilitation units on one occasion by three independent raters: two physiotherapists and one paediatric orthopaedic surgeon. All raters had many years of experience working with children with cerebral palsy, but only one of the physiotherapists had previous experience of the PPAS. The other two raters got brief instructions before assessing the children. The children were instructed by one of the physiotherapists to get into and out of supine, prone, sitting positions on a plinth and into and out of a standing position. If they were unable to do this by themselves, they were placed in the position and instructed or guided according to their cognitive abilities to maintain position, initiate flexion of the trunk (in supine) or extension (in prone), transfer weight laterally and regain position, and move out of position, according to the levels of the PPAS. If needed, children were provided with manual support to stay in position. The children were also instructed to sit, stand or lie down in prone or supine as straight as possible, or were placed as straight as possible in the specified position and allowed to settle. The experienced physiotherapist gave instructions and handled the children, and the other two raters observed. All three raters recorded their observations simultaneously and independently on separate scoring sheets. All assessments took less than 10 minutes to complete for each child.

Statistical analyses

Construct validity was evaluated for known-groups validity based on the GMFCS levels using Jonckheere-Terpstra for analysis of arithmetic average values given by the raters. Inter-rater reliability for three independent raters was calculated using weighted Kappa scores[19] with 95% non-parametric bootstrap confidence intervals calculated based on 1000 re-samples.[20,21] The levels of agreement were set to poor ($\leqslant 0.40$), fair to good (0.40–0.75), and excellent agreement ($\geqslant 0.75$).[22] The internal consistency was evaluated through Cronbach's alpha,[23] a measure of item inter-relatedness calculated with averaged values for the three raters, and corrected Item–total correlation,[24] indicating the correlation between each item and the total score. Cronbach's alpha, if item is deleted, corresponds to the value achieved if a specific item is removed and the level should exceed 0.2.[24] For all statistical computing an R software environment was used.

Results

In total 29 children with cerebral palsy (15 boys, 14 girls), born 1997–2007, median age 12 years (6–16 years) were assessed. Their gross motor function was classified as GMFCS levels II ($n = 10$), III ($n = 7$), IV ($n = 6$) and V ($n = 6$).

Distribution of scores for all raters varied between each GMFCS level in all four positions (Figure 1). The median score was higher in supine or prone positions, which require less postural ability, compared with a sitting or standing position (Table 1). The PPAS showed construct validity based on the ability of the assessment tool to differ between known groups represented by GMFCS levels II–V, where children at GMFCS level II present higher scores than children with lower levels of motor function (Table 1, Figure 1). It could differ in postural ability between individuals at different levels of gross motor function and was able to identify postural asymmetries in children at all the GMFCS levels II–V. There were no differences in scores for posture and postural ability related to the age of the children.

The PPAS showed excellent inter-rater reliability for three independent raters with weighted Kappa values of 0.77–0.99 (95% CI 0.60–1.0) (Table 2). There was a high internal consistency for all items where Cronbach's alpha if item deleted ranged from

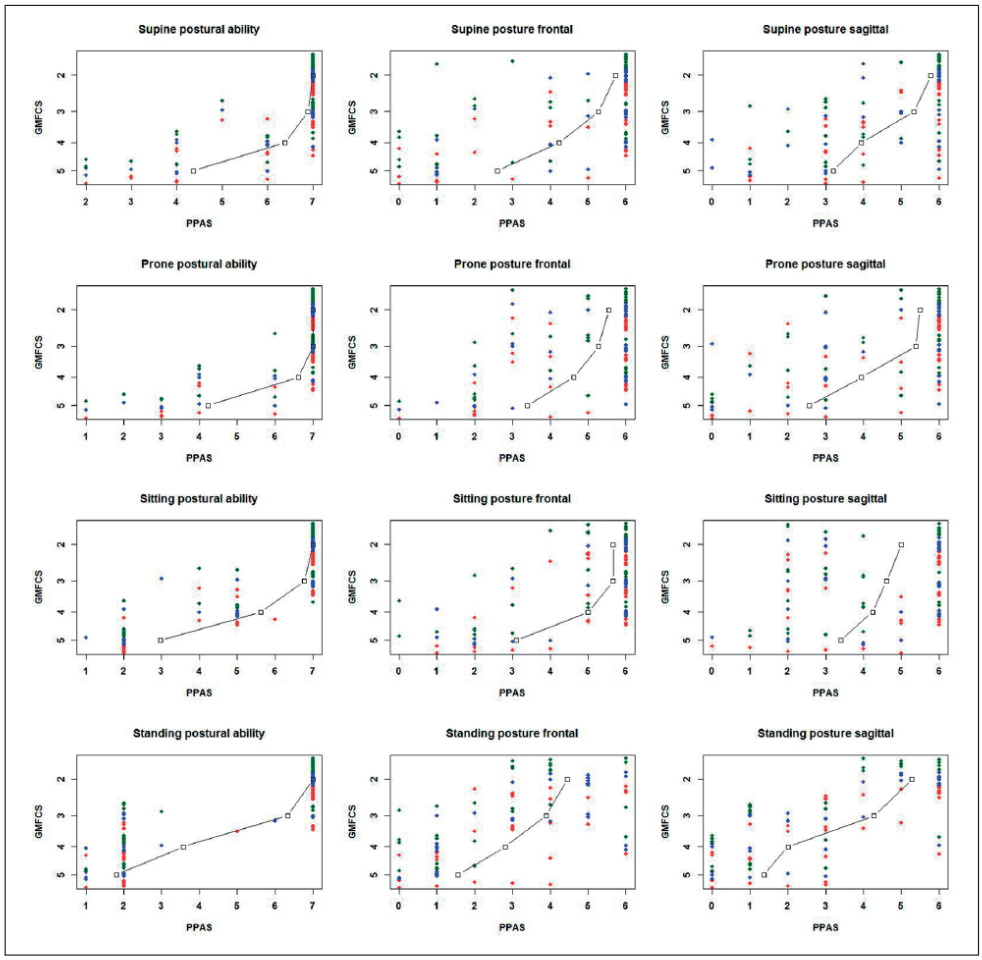


Figure 1. Distribution of scores for PPAS at each GMFCS level in all four positions.
All observations are marked with a different colour for each rater, red = rater A, blue = rater B, green = rater C. The squared points connected with a line are means of each GMFCS level.
GMFCS: Gross Motor Function Classification System; PPAS: Posture and Postural Ability Scale.

0.95–0.96 with a 95% confidence interval (CI) of 0.90–0.98 for all items. Corrected item-total correlation varied between 0.55–0.91 (95% CI 0.20–0.95) (Table 3).

Discussion

The PPAS shows sound psychometric properties for children and adolescents with cerebral palsy, comparable with a previous study with adults.[10]

There are several limitations to this study. One of the raters had special training and long experience using the PPAS, while the other raters had many years of clinical experience working with children with disabilities but no previous experience or knowledge of rating posture or postural ability. All three raters observed the children at the same time, but the children were only instructed and handled by the experienced physiotherapist. This may have affected the outcome for raters with

Table 1. Known-groups validity of the PPAS. Median, minimum and maximum values for each level of the GMFCS level II to V, and *p*-values calculated with Jonckheere-Terpstra for averaged values for the three raters.

		GMFCS II			GMFCS III			GMFCS IV			GMFCS V			*P*-value
		Median	Min	Max	Median	Min	Max	Median	Min	Max	Median	Min	Max	
Supine	Postural ability	7	7	7	7	5	7	6	4	7	3.5	2	6	<0.001
	Posture frontal	6	1	6	5	2	6	5	0	6	1	0	5	0.001
	Posture sagittal	6	4	6	4	1	6	4	0	6	3	0	6	<0.001
Prone	Postural ability	7	7	7	7	6	7	6	4	7	3	1	6	<0.001
	Posture frontal	6	3	6	5	2	6	6	2	6	2	0	6	0.003
	Posture sagittal	6	2	6	4	0	6	3	1	6	1.5	0	6	0.001
Sitting	Postural ability	7	7	7	7	3	7	5	2	7	2	1	2	<0.001
	Posture frontal	6	4	6	6	2	6	6	0	6	2	0	4	0.002
	Posture sagittal	6	2	6	4	2	6	5.5	2	6	2	0	5	0.009
Standing	Postural ability	7	7	7	5	2	7	2	1	3	2	1	2	<0.001
	Posture frontal	4	2	6	3	0	6	1	0	6	1	0	4	<0.001
	Posture sagittal	6	3	6	2	1	5	1	0	6	1	0	3	<0.001

GMFCS: Gross Motor Function Classification System.

Table 2. Weighted kappa scores for the PPAS. Inter-rater reliability for three raters calculated with weighted Kappa scores, and non-parametric bootstrap confidence intervals (95% CI).

		Weighted Kappa	95%	CI
Supine	Postural ability	0.99	0.96	1.00
	Posture frontal	0.86	0.74	0.94
	Posture sagittal	0.90	0.84	0.95
Prone	Postural ability	0.98	0.94	1.00
	Posture frontal	0.94	0.88	0.98
	Posture sagittal	0.93	0.83	0.97
Sitting	Postural ability	0.97	0.89	0.99
	Posture frontal	0.85	0.73	0.93
	Posture sagittal	0.82	0.66	0.90
Standing	Postural ability	0.97	0.96	1.00
	Posture frontal	0.77	0.60	0.88
	Posture sagittal	0.87	0.78	0.94

different professions and varying previous knowledge of using the PPAS. The weighted kappa coefficient was 0.77–0.99 indicating an excellent inter-rater reliability, in agreement with results previously reported for experienced raters (0.85–0.99).[10] From our experience, we would recommend some training to minimize errors and make the assessment smoother. Securing reproducible measures is important for any assessment tool. This could be evaluated either by repeated measures on different occasions or by different raters on the same occasion.[24] We chose to evaluate agreement between raters on the same occasion. The reason for that is that posture in children with cerebral palsy may change over time and any disagreement between two occasions could represent responsiveness to change rather than measurement error. In the previous evaluation of the PPAS,[10] the ratings were based on photos and videos, however the present study shows similar results in spite of different

Table 3. Internal consistency of the PPAS. Cronbach's alpha if item deleted with 95% CI for three independent raters followed by corrected item-total correlation with 95% CI showing the correlation between each item and the total score.

		Cronbach's α	95%	CI	Item-total	95%	CI
Supine	Postural ability	0.95	0.91	0.97	0.84	0.68	0.92
	Posture frontal	0.95	0.91	0.97	0.83	0.56	0.92
	Posture sagittal	0.95	0.91	0.97	0.79	0.49	0.91
Prone	Postural ability	0.95	0.91	0.97	0.85	0.72	0.93
	Posture frontal	0.95	0.91	0.97	0.78	0.40	0.89
	Posture sagittal	0.95	0.91	0.97	0.82	0.55	0.91
Sitting	Postural ability	0.95	0.90	0.97	0.91	0.82	0.95
	Posture frontal	0.95	0.91	0.97	0.87	0.73	0.94
	Posture sagittal	0.96	0.92	0.98	0.55	0.20	0.79
Standing	Postural ability	0.96	0.92	0.97	0.70	0.44	0.83
	Posture frontal	0.95	0.91	0.97	0.77	0.58	0.87
	Posture sagittal	0.95	0.91	0.97	0.72	0.50	0.83

methodology. The numbers are quite small, particularly for the children with more severe impairments, with a total of 12 children at GMFCS level IV–V compared with a total of 17 children at GMFCS II–III. However, the results are statistically significant, but a bigger sample might have provided a narrower confidence interval.

The internal consistency represents the average of the correlations among all items. It was 0.95–0.96, which by far exceeds the recommended 0.8.[24] We anticipated a high homogeneity since all items assess aspects of posture and postural ability. For the same reason, methods such as factor analysis, often used to differentiate between items in different domains in questionnaires, would not be appropriate in this case. Corrected item-total correlation showed a slightly lower value for sitting posture in the sagittal view. This is an important consideration when using the PPAS in clinical practice. It can be difficult to assess whether or not the hips are flexed to approximately 90° depending on the position of the pelvis and the height of the plinth. In sitting, if the plinth is not adjustable or if using a chair, provision of additional support for the feet is necessary, especially for children at different heights. The results are comparable with the findings of a similar previous study using the PPAS to assess posture in adults with cerebral palsy.

Construct validity of the PPAS was evaluated through its ability to differ between known groups in terms of the GMFCS levels in children with cerebral palsy. There are many tools to assess balance for individuals who are ambulant, but most of them require at least the ability to maintain sitting or standing independently. The PPAS is designed for use with people at a lower level of gross motor function. Children at GMFCS level II can walk and stand unsupported. The highest level of ability is to move into and out of position, therefore, an anticipated ceiling effect in postural ability was seen for children at GMFCS level II. The strength of the PPAS is that it identifies postural asymmetries and deviations at all GMFCS-levels presented in this study.

Children with severe motor impairments frequently remain in a sitting or lying position for several hours a day. A sustained posture over longer periods of time leads to tissue adaptation and development of secondary complications, such as contractures, deformities and pain.[2,3,25] However, this can be prevented by early detection and appropriate interventions,[6,9,26,27] including provision of adaptive seating, standing or night-time support equipment.[27–29] The PPAS is sensitive to identify small asymmetries and deviations at all levels of motor function and is likely to detect asymmetries at an early stage. It is well recognized that persistent asymmetry will increase over time, leading to

established contracture and deformity.[1–4] Early detection is essential if these problems are to be prevented or minimized.

The ability of the PPAS to identify problems of posture and postural ability at an early stage, not only highlights the need for early intervention, but provides information on what postural support is appropriate and where it needs to be applied. For example, children rated as level 1 (unplaceable) would require customized seating and standing support owing to fixed deformities and contractures. The quality of posture indicates if support or adaptations are required to improve weight distribution, or to get head, trunk, pelvis, legs, arms and feet in a neutral position. In addition, the assessment does not require any special equipment; it is easy to use in a clinical setting and takes about 10 minutes to complete. Its use should facilitate evaluation of those therapeutic interventions designed to increase functional ability and to prevent secondary complications.

The PPAS shows construct validity, internal consistency and excellent inter-rater reliability for raters with experience of children with cerebral palsy. It can detect postural deficits and asymmetries, which enable early detection of potential problems and provides information relevant to postural support solutions in order to improve function and prevent musculoskeletal deformities.

Clinical messages

- The Posture and Postural Ability Scale shows high psychometric properties for children with cerebral palsy.
- The Posture and Postural Ability Scale identifies asymmetries in children at varying levels of motor function and can be used for children with mild to severe postural deficits.

Acknowledgements

The authors would like to thank all the children and families for participating in this study, the physiotherapists in Eslöv, Helsingborg, Lund, Malmö and Trelleborg for assistance, and especially Anette Sällvik for assessing all children. We would also like to express our deep gratitude to Pauline Pope for valuable input and for proof reading and improving the manuscript.

Conflict of interest

The authors declare that they have no conflict of interests.

Contributors

ERB designed the study, examined the children, collected and analysed the data, and drafted the manuscript. MPB recruited and examined the children, analysed the data, improved and revised the manuscript. TC analysed the data, performed all statistical analyses, improved and revised the manuscript. All authors approved the final draft.

Funding

We would like to thank the Centre for Clinical Research Västerås, the Faculty of Medicine, Lund University and Stiftelsen för bistånd åt rörelsehindrade i Skåne, for financial support.

References

1. Brown K. Positional deformity in children with cerebral palsy. *Physiother Theory Practice* 1985; 1: 37–41.
2. Fulford FE and Brown JK. Position as a cause of deformity in children with cerebral palsy. *Dev Med Child Neurol* 1976; 18: 305–314.
3. Porter D, Michael S and Kirkwood C. Is there a relationship between preferred posture and positioning in early life and the direction of subsequent asymmetrical postural deformity in non ambulant people with cerebral palsy? *Child Care Health Dev* 2008; 34: 635–641.
4. Rodby-Bousquet E, Czuba T, Hägglund G and Westbom L. Postural asymmetries in young adults with cerebral palsy. *Dev Med Child Neurol* 2013; 55: 1009–1015.
5. Hägglund G, Alriksson-Schmidt A, Lauge-Pedersen H, Rodby-Bousquet E, Wagner P and Westbom L. Prevention of dislocation of the hip in children with cerebral palsy: 20-year results of a population-based prevention programme. *Bone Joint J* 2014; 96-B: 1546–1552.
6. Hägglund G, Andersson S, Duppe H, Lauge-Pedersen H, Nordmark E and Westbom L. Prevention of severe contractures might replace multilevel surgery in cerebral palsy: results of a population-based health care programme and new techniques to reduce spasticity. *J Pediatr Orthop B* 2005; 14: 269–273.
7. Novak I. Stand up and be counted. *Dev Med Child Neurol* 2013; 55: 974.
8. Persson-Bunke M, Hägglund G and Lauge-Pedersen H. Windswept hip deformity in children with cerebral palsy. *J Pediatr Orthop B* 2006; 15: 335–338.

9. Persson-Bunke M, Hägglund G, Lauge-Pedersen H, Wagner P and Westbom L. Scoliosis in a total population of children with cerebral palsy. *Spine* 2012; 37: E708–713.
10. Rodby-Bousquet E, Agustsson A, Jonsdottir G, Czuba T, Johansson AC and Hagglund G. Interrater reliability and construct validity of the Posture and Postural Ability Scale in adults with cerebral palsy in supine, prone, sitting and standing positions. *Clin Rehabil* 2014; 28: 82–90.
11. Bouisset S and Do MC. Posture, dynamic stability, and voluntary movement. *Neurophysiol Clin* 2008; 38: 345–362.
12. Pope PM. *Severe and complex neurological disability: Management of the physical condition*. Edinburgh: Butterworth-Heinemann/Elsevier, 2007.
13. Hallett R, Hare N and Milner A. Description and evaluation of an assessment form *Physiotherapy* 1987; 73: 220–225.
14. Green EM and Nelham RL. Development of sitting ability, assessment of children with a motor handicap and prescription of appropriate seating systems. *Prosthet Orthot Int* 1991; 15: 203–216.
15. Pountney TE, Cheek L, Green E, Mulcahy C and Nelham R. Content and criterion validation of the chailey levels of ability. *Physiotherapy* 1999; 85: 410–416.
16. Westbom L, Hägglund G and Nordmark E. Cerebral palsy in a total population of 4–11 year olds in southern Sweden. Prevalence and distribution according to different CP classification systems. *BMC Pediatr* 2007; 7: 41.
17. (SCPE) SoCPiE. Surveillance of cerebral palsy in Europe: A collaboration of cerebral palsy surveys and registers. *Dev Med Child Neurol* 2000; 42: 816–824.
18. Palisano RJ, Rosenbaum P, Bartlett D and Livingston MH. Content validity of the expanded and revised Gross Motor Function Classification System. *Dev Med Child Neurol* 2008; 50: 744–750.
19. Conger AJ. Integration and generalization of kappas for multiple raters. *Psychological Bulletin* 1980; 88: 322–328.
20. Davison AC and Hinkley DV. *Bootstrap methods and their application*, Chapter 5 Confidence Intervals: 191–251. Cambridge: Cambridge University Press, 1997.
21. DiCiccio TJ and Efron B. Bootstrap confidence intervals. *Statistical Science* 1996; 11: 189–212.
22. Fleiss JL. *Statistical methods for rates and proportions*. 2nd ed. New York: Wiley, John and Sons, Incorporated, 1981, p. 321.
23. Cortina JM. What is coefficient alpha? An examination of theory and applications. *J App Psychol* 1993; 78: 98–104.
24. Streiner DL and Norman GR. *Health measurement scales: A practical guide to their development and use*. Oxford: Oxford University Press, 2008.
25. Saito N, Ebara S, Ohotsuka K, Kumeta H and Takaoka K. Natural history of scoliosis in spastic cerebral palsy. *Lancet* 1998; 351: 1687–1692.
26. Hägglund G, Andersson S, Duppe H, Lauge-Pedersen H, Nordmark E and Westbom L. Prevention of dislocation of the hip in children with cerebral palsy. The first ten years of a population-based prevention programme. *J Bone Joint Surg Br* 2005; 87: 95–101.
27. Pountney T, Mandy A, Green E and Gard P. Management of hip dislocation with postural management. *Child Care Health Dev* 2002; 28: 179–185.
28. Rodby-Bousquet E and Hägglund G. Sitting and standing performance in a total population of children with cerebral palsy: A cross-sectional study. *BMC Musculoskelet Disord* 2010; 11: 131.
29. Pope PM, Bowes CE and Booth E. Postural control in sitting the SAM system: Evaluation of use over three years. *Dev Med Child Neurol* 1994; 36: 241–252.