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Posture and Mobility of Children with Cerebral Palsy

Jackie Casey



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DOCTORAL DISSERTATION

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To be defended at föreläsningssal 3 i Blocket, Lund.

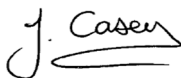
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Title and subtitle: Posture and mobility of children with cerebral palsy		
Abstract <i>Background:</i> Cerebral palsy (CP) is the most common cause of motor disability in childhood primarily affecting posture and mobility. Secondary complications such as contractures and deformities may affect their seating or lying posture, their ability to move around independently and cause pain. All of which can impact upon the child's ability to participate and engage in everyday activities and their quality of life. <i>Purpose:</i> To increase knowledge of the prevalence and associations between postural asymmetries, scoliosis, windswept hip deformity, contractures and pain of children with CP in supine and sitting; the time sequence and incidence of scoliosis and windswept hip deformity and to identify risk factors preventing independent wheelchair mobility. <i>Methods:</i> All studies were based on data from the Swedish national registry and follow-up program for children with CP at all levels of the Gross Motor Function Classification System (GMFCS) levels I-V. <i>Study I-III</i> were cross-sectional studies. <i>Study I</i> examined wheelchair mobility in children aged 0-11 and analysed if reduced hand function or range of motion were risk factors for not self-propelling or driving their wheelchair. <i>Study II-III</i> included children 0-18 years, and <i>Study II</i> focused on postural asymmetries, postural ability and pain in children 0-18 years; whilst <i>Study III</i> analysed associations between posture, deformities and contractures. <i>Study IV</i> was a longitudinal cohort study of 4148 children followed until they developed either scoliosis or windswept hip deformity. <i>Results:</i> Only 1 in 10 children self-propelled a manual wheelchair independently, whilst 3 in 4 children could independently drive a power wheelchair. Poor hand function or gross motor function were the greatest risk factors for being unable to self-propel or drive a wheelchair. Over half the children had postural asymmetries in sitting or supine positions; 10.5% had scoliosis, 8.7% windswept hips, while 19.2% had knee contractures and 6.6% hip flexion contractures. Postural asymmetries increased with age and GMFCS levels. An asymmetric posture doubled the likelihood of pain; whilst severe postural asymmetries increased the likelihood of scoliosis (OR 9.1), windswept hip deformity (OR 5.7-8.8), hip (OR 6.7) and knee flexion contractures (OR 12.2). Children unable to maintain or change position were more likely to have postural asymmetries in supine (OR 2.6-7.8) or in sitting (OR 1.5-4.2). The likelihood of pain increased if the child had hip flexion contractures (OR 1.5) or windswept hip deformity (OR 1.6). There was a higher incidence of windswept hip deformity (16.6%) than scoliosis (8.1%) as the first deformity; windswept hip deformity developed first in more children at higher GMFCS levels, and with dyskinetic (20.0%), or spastic CP (17.0%), whereas scoliosis developed in more children with GMFCS level V (19.8%) or dyskinetic CP (17.9%). <i>Conclusion:</i> Most children aged 0-11 years could not self-propel their manual wheelchair; whilst more children could independently drive a power wheelchair. Power mobility should be considered earlier. Windswept hip deformity presents earlier than scoliosis. Postural asymmetries are associated with inability to change position in supine and sitting; pain; and with having scoliosis, windswept hip deformity, hip and knee flexion contractures. It is important to reduce postural asymmetries for children with CP to reduce the risk of having scoliosis, windswept hip deformity, contractures and pain.		
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Jackie Casey



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Cover photo by Elisabet Rodby-Bousquet, “*Balancing the elements - body segments*”

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*“Our prime purpose in this life is to help others. And if you
can't help them, at least don't hurt them.”
- Dalai Lama XIX*

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Original papers

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

- I. Rodby-Bousquet E, Paleg G, Casey J, Wizert A & Livingstone R. Physical risk factors influencing wheeled mobility in children with cerebral palsy: a cross-sectional study. *BMC Pediatrics*, 2016, DOI 10.1186/s12887-016-0707-6
- II. Casey J, Rosenblad A & Rodby-Bousquet E. Postural asymmetries, pain, and ability to change position of children with cerebral palsy in sitting and supine: a cross-sectional study. *Disability and Rehabilitation*, 2020; Nov 3: 1-9. DOI 10.1080/09638288.2020.1834628
- III. Casey J, Agustsson A, Rosenblad A & Rodby-Bousquet E. Relationship between scoliosis, windswept hips and contractures with pain and asymmetries in sitting and supine in 2,450 children with cerebral palsy. *Disability & Rehabilitation*, 2021; DOI: 10.1080/09638288.2021.1971308
- IV. Casey J, Rosenblad A, Agustsson A, Lauge-Pedersen H & Rodby-Bousquet E. Incidence of scoliosis and windswept hip deformity: which comes first in 4148 children with cerebral palsy? A longitudinal cohort study. Manuscript in review Feb. 2022

Abbreviations

CI	Confidence Intervals
CP	Cerebral Palsy
CPUP	Cerebral Palsy Follow-up Programme and National Quality Registry
GMFCS	Gross Motor Function Classification System
HR	Hazard Ratio
MACS	Manual Ability Classification System
MP	Migration Percentage
OR	Odds Ratio
PO	Pelvic Obliquity
PPAS	Posture and Postural Ability Scale
RI	Reimer's Index
ROM	Range of Motion
SCPE	Surveillance of Cerebral Palsy in Europe
SD	Standard Deviation
WSH	Windswept hip deformity

Definitions

Cerebral palsy	An umbrella term (1) describing a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances of sensation, perception, cognition, communication, and behaviour, by epilepsy and by secondary musculoskeletal problems (2).
Contracture	Limitation in the passive range of motion of a joint, secondary to tissue adaptation, shortening of the connective tissues and muscles (3).
Hip dislocation	Localisation of the femoral head outside of the acetabulum socket, with a Reimer's migration percentage of 100% (4).
Hip flexion contracture	Contracture of the hip in a flexed position preventing extension of the hip to neutral (0°).
Knee flexion contracture	Contracture of the knee in a flexed position preventing extension of the knee to neutral (0°).
Posture	Anatomical alignment of body segments in relation to each other, and to the support surface (5, 6).
Postural ability	An individual's ability to stabilize their body segments in relation to both each other and to the supporting surface during static and dynamic conditions; the ability to maintain and or change position to participate in an activity (5, 6).
Scoliosis	A lateral deviation of the spine in the frontal plane of at least 10° (7).
Windswept hip deformity	A hip deformity calculated based upon passive hip range of motion of abduction, internal and external rotation with a difference of >50% between the left and the right side (8).

The thesis at a glance

Study	Questions	Methods	Results	Conclusions
I	<p>How many children have independent mobility in their wheelchairs indoors and outdoors?</p> <p>Does hand function and upper extremity range of movement affect independent wheeled mobility?</p>	Cross-sectional study of 2,328 children with CP 0-11 years.	<p>Indoors: 28% self-propelled their manual; 66% drove power independently.</p> <p>Outdoors: 10% self-propelled their manual; 75% drove power independently.</p> <p>Poor hand function (MACS IV-V) and gross motor function (GMFCS V) are the greatest risk factors for not driving power independently.</p>	Most children had a manual wheelchair. Most children could not self-propel a manual wheelchair. Power mobility should be considered earlier as an option for independent wheeled mobility.
II	<p>What are the associations between postural asymmetries, postural ability, and pain in sitting and supine positions?</p>	Cross-sectional study of 2,735 children with CP 0-18 years.	<p>Postural asymmetries occurred for 60.2% in sitting, and 53.6% in supine, and across all age groups. Children with severe asymmetric postures are twice as likely to have pain. Inability to maintain or change position increases the risk for postural asymmetries in supine (OR 2.6-7.8) and sitting (OR 1.5-4.2).</p>	Asymmetric postures are associated with pain and inability to change position in sitting and supine position. There is a need to assess posture and intervene earlier to address asymmetric posture.
III	<p>What is the prevalence of spine, hip and knee deformities and contractures?</p> <p>What are the associations between postural asymmetries, ability to change position, and pain?</p>	Cross-sectional study of 2,450 children with CP 0-18 years.	<p>In total 10.5% had scoliosis, 8.7% windswept hips, 6.6% hip and 19.2% knee contracture, 1% <90° hip contracture, and 0.2% hip dislocation.</p> <p>Severe postural asymmetries increased the likelihood for scoliosis 9 times, and 6-9 times for windswept hip deformity (WSH).</p> <p>Hip flexion contractures and WSH increased the likelihood for pain by 1.5-1.6 times.</p>	Scoliosis, windswept hips and hip-, knee contractures are associated with postural asymmetry. Pain is associated with windswept hip or hip contractures. Reducing postural asymmetries may reduce the risk of pain.
IV	<p>Which deformity occurs first, scoliosis or windswept hip deformity?</p> <p>Is there a difference in the incidence of scoliosis and windswept hip deformity for children at different GMFCS-levels and subtypes?</p>	Longitudinal cohort study of 4,148 children with CP born 1990 to 2018, reported in the national registry before 6 th birthday.	<p>Most children do not develop either deformity.</p> <p>WSH develops first most often (16.6%), whilst scoliosis develops first for 8.1% children.</p> <p>Children with spasticity are more likely to develop WSH first (17%). Children with dyskinesia are more likely to develop either WSH (20%), or scoliosis (17.9%), and to do so at a younger age.</p>	WSH develops first more often than scoliosis. Children with dyskinesia and those with GMFCS level IV and V have a higher incidence of scoliosis and WSH and should have earlier targeted interventions.

Why it matters

In my clinical practice I have the privilege of working with children, young people and adults with cerebral palsy who have complex postural needs and who require support in sitting or with wheelchair mobility. From working with my clients and their caregivers I have seen first-hand how some of postural asymmetries, deformities and contractures can impact directly upon their ability to lie, sit, and to engage and participate in everyday activities.

As clinicians we believe that good postural support can offer comfort, maintain postural alignment, and provide core stability in sitting so that the young person can engage or participate in activities of daily living using the skills they have available to them.

However, despite our efforts some of our clients still develop postural asymmetries, contractures, and deformities.



One of my first clients, let's call her Ruth, was 17 years old and had cerebral palsy, GMFCS and MACS levels IV. She had outgrown her current manual and power wheelchair seating systems. The custom moulded seating no longer matched her body contours, was uncomfortable, and even painful to sit in for prolonged periods of time. On observation she was presenting with postural asymmetries in sitting, which on physical examination included knee flexion contractures, bilateral limited hip flexion, right windswept hip deformity, a scoliosis with left side convexity, and a lumbar lordosis. Ruth was also struggling to self-propel her wheelchair for any distance at all, which had left her dependent upon her mother for mobility within her home and community environments. Subsequently she spent less time socialising with her peers outside of the classroom.

Further, it was becoming increasingly difficult to provide suitable seating without using ever more tilt within her wheelchair seating, creating more compromises around shear on her buttocks and foot support positioning.

Ruth's story clearly illustrates the direct impact that her postural presentation, deformities, and contractures have upon her ability to engage and participate in everyday life activities, to socialise with other young people her own age, and to be independent.

Reflecting upon her clinical journey, I wanted to learn more as to why had she developed deformities and contractures? Was she typical of a young person with cerebral palsy regarding her postural asymmetries? What were the risk factors restricting her from being able to self-propel or drive her wheelchairs? Which deformity occurred first, or should we have addressed? How many children experience pain and is it linked to her posture or deformities presentation? So many questions needing answers so that I could understand her journey and those of other children with cerebral palsy. Additionally, could I recognise postural patterns to better inform my clinical practice with these children and their families to reduce the risk of these postural asymmetries, contractures and deformities occurring?

Introduction

Cerebral palsy

Different definitions and classifications have been used to describe cerebral palsy (CP) over the decades. However, as early as 1843 Dr William John Little, an English orthopaedic surgeon first presented his influential work on contractures and deformities seen in children through a series of public lectures (9). He described the cause of the spasticity and paralysis of CP as being a result of brain damage occurring at birth. He identified two primary types of deformities; one being congenital in nature, and the other type of deformities resultant from birth difficulties as ‘spastic rigidity’ (10). In his published work Little described the spastic deformities of 47 cases of children with CP into hemiplegic rigidity (affecting one side more than the other), paraplegia (affecting both legs more than the arms), and generalised rigidity. At the time this spastic rigidity was known as Little’s disease but is now known to be synonymous with cerebral palsy (9).

However, in 1893 Sigmund Freud, neuropathologist, maintained that CP should be classified using only clinical findings (9). Freud advocated that CP could also be caused early during the pregnancy and not solely at birth. He went on to be the first to use the term ‘cerebral diplegia’ in children to define all types of bilateral CP (11).

Developments in assessment methods and increased access to medical diagnostic technologies has led to increases in knowledge (12) and understanding of CP and appreciation of both its complexity of presentation and impact upon the individual and their everyday life. This has resulted in changes in the definition of CP, acknowledging this increased appreciation.

Mutch et al. (1) described CP as “an umbrella term covering a group of non-progressive, but often changing, motor impairment syndromes, secondary to lesions or anomalies of the brain arising in early stages of its development”. This definition of CP was superseded in 2007 when an international group of experts met to reappraise and update the definition and classification of CP. This group of experts defined 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 foetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication, and behaviour, by epilepsy, and by secondary musculoskeletal problems” (2).

Despite being described as non-progressive there are musculoskeletal changes/manifestations that happen over time. These can include joint range of motion limitations, contractures, and deformities. We now have the added recognition of the International Classification of Functioning, Disability, and Health (ICF) framework acknowledging how these elements of the body structures and function domain directly influence activity and participation domains, and overall quality of life for the individual (13).

Prevalence of CP

CP is the most common childhood motor disability (14, 15), with a prevalence of approximately 1.96 per 1,000 live births in Sweden (16), whilst the prevalence in Europe is 1.77 per 1,000 live births (17). The overall prevalence of cerebral palsy worldwide appears to be reducing (18). The reasons for this reduction are likely to be multifaceted, with greater implementation of preventative and peri- and post-natal interventions. Such interventions include improved access to obstetric care and nutrition during the pregnancy, changes in obstetric care (16, 19), heightened awareness of risk factors associated with CP, and the use of therapeutic hypothermia (17, 19).

Criteria for CP

CP is a diverse disorder in terms of its aetiology, motor presentation, severity of limitations and impairments (12). Over the last couple of centuries there have been many attempts to classify CP into different subtypes; with some based on neurological presentation and others upon laterality of body involved. The classification system by the Surveillance of Cerebral Palsy in Europe network (SCPE) has been universally adopted since 2000 (14) by registers, databases and clinicians alike. This common terminology enables clearer communication between the healthcare professionals, researchers, and families. CP is classified into different categories and sub-categories or sub-types, including ataxic, dyskinetic, spastic and non-classified/ mixed type. The SCPE also developed a decision tree summarising the inclusion and exclusion eligibility criteria for cerebral palsy (Figure 1). These criteria help to identify children who meet the definition of CP, with their diagnosis based only on clinical description of their presentation. The SCPE eligibility and classification systems have been used in papers I-IV.



Decision tree for Cerebral Palsy

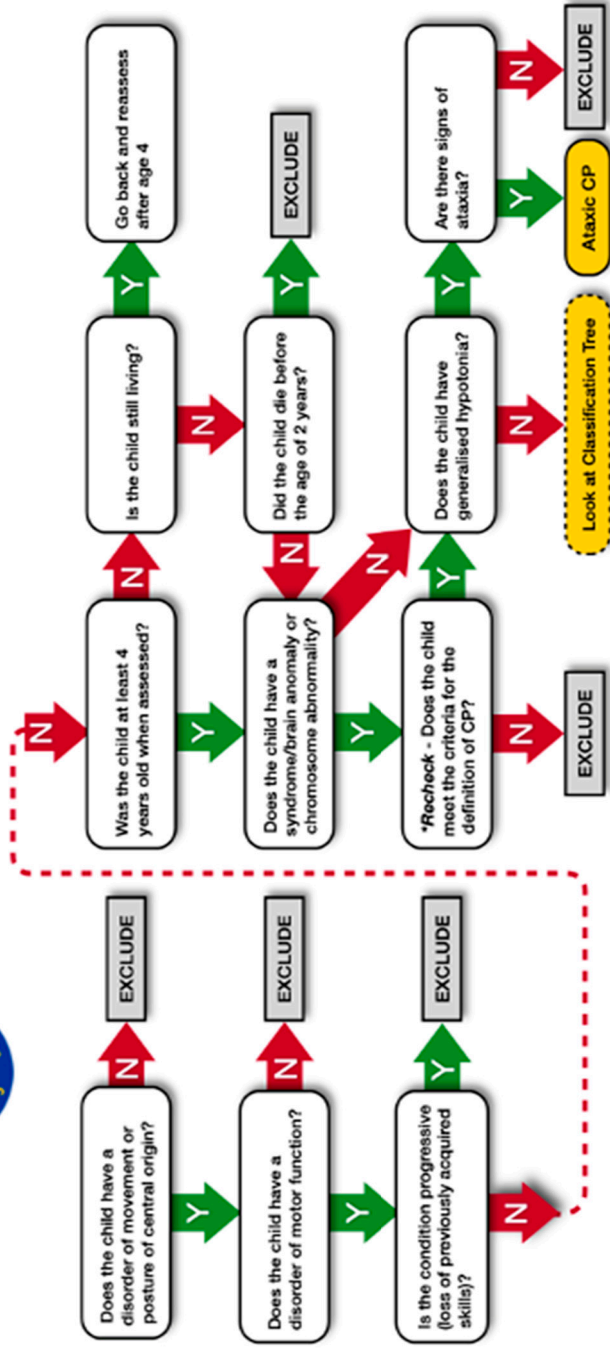


Figure 1. The SCPE Decision tree for Cerebral Palsy. This figure is reprinted with permission from Mac Keith Press (Cans 2000).

Classification of gross motor function

Gross motor function is the primary limitation in children with cerebral palsy and is universally described using the Gross Motor Function Classification System (GMFCS) Expanded and Revised (20, 21). The first version was published in 1997, and so was not available when the follow-up program was for children with CP was commenced in Sweden in 1994. The GMFCS is a five-point ordinal scale (I-V), where level I describes the highest level of gross motor function and level V the lowest (Figure 2) of children aged up to 18 years (22). It is reported as having high inter-rater reliability and validity for prediction of gross motor function (23-25), high test-retest reliability (26), and good content validity (22).

Classification of children's gross motor function using the GMFCS is considered to be relatively stable and the classification level does not usually change over time (25), and the child's level often plateaus at around 6-8 years (27). Therefore, it can be used to predict future gross motor function (26) and anticipate what mobility assistive technology support the child is likely to require. Typically, those children with GMFCS level I will not require any mobility assistive technology to walk or move; whilst those with GMFCS levels IV-V require the use of a wheelchair for all their independent mobility. The levels of the GMFCS are clearly illustrated in Figure 2 (*next page*). For those children with mobility limitations wheelchair provision is vital in providing the opportunity for self-initiated movement, exploring one's physical and social environments (28, 29), play and to develop independence and an internal locus of control. Without the experience of mobility, the child will experience deprivation of stimuli which can result in developmental delays, a learned helplessness (30) and ultimately be caught within a cycle of deprivation (31, 32) (Figure 3).

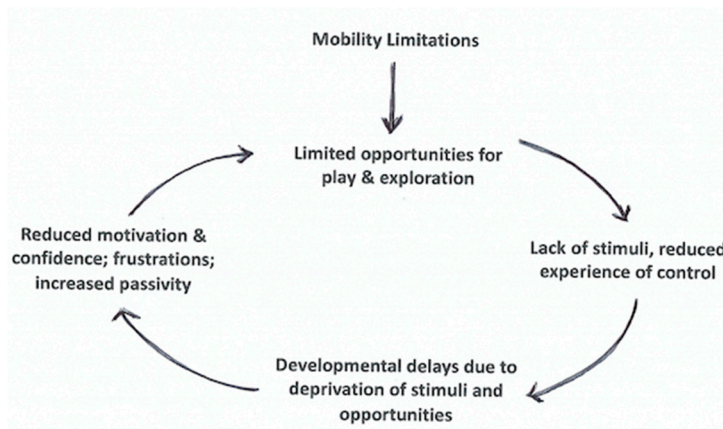
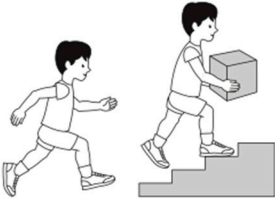
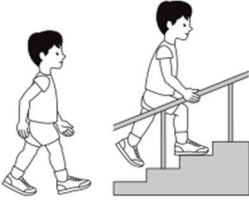
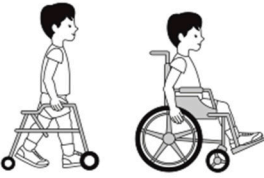




Figure 3. Nisbet et al. 1996 Cycle of deprivation

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

	<p>GMFCS Level I</p> <p>Children walk at home, school, outdoors and in the community. They can climb stairs without the use of a railing. Children perform gross motor skills such as running and jumping, but speed, balance and coordination are limited.</p>
	<p>GMFCS Level II</p> <p>Children walk in most settings and climb stairs holding onto a railing. They may experience difficulty walking long distances and balancing on uneven terrain, inclines, in crowded areas or confined spaces. Children may walk with physical assistance, a hand-held mobility device or used wheeled mobility over long distances. Children have only minimal ability to perform gross motor skills such as running and jumping.</p>
	<p>GMFCS Level III</p> <p>Children walk using a hand-held mobility device in most indoor settings. They may climb stairs holding onto a railing with supervision or assistance. Children use wheeled mobility when traveling long distances and may self-propel for shorter distances.</p>
	<p>GMFCS Level IV</p> <p>Children use methods of mobility that require physical assistance or powered mobility in most settings. They may walk for short distances at home with physical assistance or use powered mobility or a body support walker when positioned. At school, outdoors and in the community children are transported in a manual wheelchair or use powered mobility.</p>
	<p>GMFCS Level V</p> <p>Children are transported in a manual wheelchair in all settings. Children are limited in their ability to maintain antigravity head and trunk postures and control leg and arm movements.</p>

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

Illustrations Version 2 © Bill Reid, Kate Willoughby, Adrienne Harvey and Kerr Graham,
The Royal Children's Hospital Melbourne ERC151050

Figure 2. The Gross Motor Function Classification System (GMFCS E&R) for ages 6-12 years.
This figure is reprinted with permission from Professor Kerr Graham.

Classification of hand function

The ability of children with CP to use their hands individually and bilaterally to grasp and handle daily objects can be affected by upper limb spasticity, muscle weakness, or limitations in joint range of motion (ROM). The Manual Ability Classification System (MACS) is used to classify how children use their hands together to handle objects in everyday meaningful situations (25, 33, 34) and can be used to determine the need for assistance or adaptation to perform manual activities. It describes five levels of manual ability, ranging from level I, able to handle objects easily, through to level V, having severe manual ability limitations and not able to handle objects, and requires assistance to do so (35)(Table 1). The MACS is reported to have good stability over time (34, 36), and excellent inter-rater reliability between therapists, and between parents and therapists (33, 36).

The House Functional Classification describes functional grasp of each hand (Table 2), House thumb-in-palm deformity classification (Table 3) determines impact of any dynamic spasticity (37); and the Zancolli classification of finger and wrist extension for grasp and release patterns (Table 4) are used to provide an understanding of the function of each hand, and together with the MACS they provide a comprehensive picture of the overall hand function (37).

Table 1. Manual Ability Classification System (MACS) for handling objects

Level	Description
I	Handles objects easily and successfully
II	Handles most objects but with somewhat reduced quality and/ or speed of achievement
III	Handles objects with difficulty; needs help to prepare and/ or modify activities
IV	Handles a limited selection of easily managed objects in adapted situations
V	Does not handle objects and has severely limited ability to perform even simple actions

Table 2. House Functional Classification for handling objects

Group	Functional level	Description
Does not use	0	Does not use
	1	Stabilizes without grasp
Passive hand	2	Fair passive grasp
	3	Good passive grasp
	4	Poor active grasp
Active hand	5	Fair active grasp
	6	Good active grasp
Manipulating hand	7	Reduced dexterity
	8	No limitation

Table 3: House Classification of Thumb-in-palm deformity

Type	Description
I	Metacarpal adduction
II	Metacarpal adduction and MCP joint flexion
III	Metacarpal adduction and MCP joint hyperextension
IV	Metacarpal adduction and MCP and IP joint flexion

Note: MCP, metacarpophalangeal; IP, interphalangeal

Table 4. Zancolli Classification of voluntary grasp and release of the hand, wrist and fingers

Group	Degree of flexion spasticity	Description
1	Minimal	Complete extension of fingers with neutral wrist position, or with <20° flexion
2	Moderate	Fingers can be actively extended but only with >20° wrist flexion
Subgroup 2a		Active extension of the wrist with fingers flexed; Finger extension with >20° wrist flexion
Subgroup 2b		No active wrist extension even with fingers flexed; Finger extension with >20° wrist flexion
3	Severe	Unable to extend fingers, even with maximal wrist flexion

Posture and postural ability

Although posture does not have a universal definition it is frequently associated with having a symmetrical appearance. However, throughout this thesis the use of posture refers to the alignment of the body segments both in relation to each other and to their support surface (6).

Postural ability is used to refer to the ability of the individual to maintain balance and stability of these body segments under both static and dynamic conditions; as well as the ability to maintain and or change a basic body position to engage in an activity (6, 38, 39).

Seminal work by Fulford and Brown (40) described positional deformities and highlighted the effect of prolonged time spent in one position upon the development of postural asymmetries and deformities for the immobile child with CP. Further, this inability to move or change position, in combination with prolonged periods of immobility in an asymmetric position has been recognised as contributing to the development of postural asymmetries (6, 41), tissue adaption (41) and joint contractures (42, 43). The effect of this combination is further intensified by the effects of gravity and ground reaction forces from the support surface upon the musculoskeletal system. This gives rise to the concept known as the ‘Human Sandwich’ (44), whereby the individual becomes the filling being both held in place, and simultaneously compressed between these forces (45) (Figure 4).

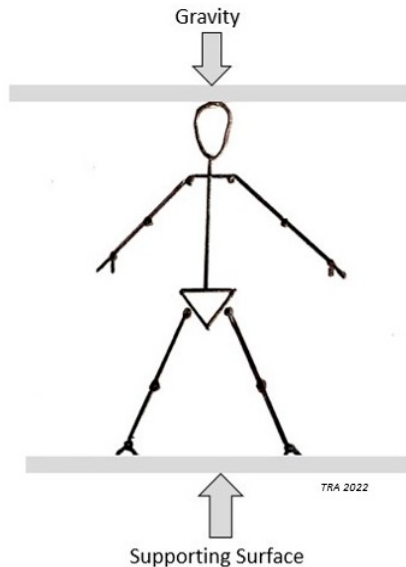


Figure 4. The human sandwich as described by Noreen Hare in 1987

Assessment of posture and postural ability

The Posture and Postural Ability Scale (PPAS) was developed by Pope and colleagues (5) and designed to be used with all persons with complex physical needs. The PPAS offers an objective measure of the individual's postural ability to maintain and change position, and their postural symmetry from sagittal and frontal planes to be recorded, for sitting, standing, supine lying and prone positions. It has been reported to have good internal consistency, and excellent inter-rater reliability (weighted kappa score = 0.77-0.99) and construct validity for both children and adults with CP (5, 46).

This tool can be utilised to justify the provision of specialised seating and positioning support providing measurement of how the assistive devices can support and promote postural alignment. In addition, the PPAS can measure the effectiveness of providing appropriate support or assistive technology to maintain a functional symmetric posture. A copy of PPAS for sitting and supine can be found in the Appendix 1 and scoring in Table 5, as well as an example of a child in supine with postural asymmetries in Figure 5.

Table 5. PPAS Scoring tables for supine and sitting. This table is reprinted with permission from Dr. Rodby-Bousquet.

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 (Yes = 1 point, No = 0 points)		
Frontal View	Sagittal View	Sagittal View
Sitting, Supine	Sitting	Supine
Head midline	Head midline	Head midline
Trunk symmetrical	Trunk in neutral position	Trunk in neutral position
Pelvis neutral	Pelvis neutral	Pelvis neutral
Legs separated and straight relative to pelvis	Hips mid-position (90°)	Legs straight, hips/ knees extended
Arms resting by side	Knees mid-position (90°)	Feet resting in normal position
Weight evenly distributed	Feet mid-position/ flat on floor	Weight evenly distributed



Figure 5. Illustration showing child's postural alignment in supine shows asymmetries in frontal and sagittal views.

Deformities of the spine and hips

Children with CP are typically born without musculoskeletal deformities and with a normal pelvis, hips, and spine alignment (47, 48). For some children there may be a postural mal-alignment preference starting to prevail, or a preferred posture presenting (41). At this early-stage children's anatomical alignment is flexible and correctable towards the midline. However, after birth, several factors may influence postural and skeletal alignment in sitting, lying, and standing, such as muscle weakness, imbalance, lack of weight-bearing, or spasticity.

The inability to move or change position, combined with spending long periods of time in an asymmetric position has also been attributed to the development of a positional deformity (49) and the development of deformities and contractures (41, 50).

Scoliosis and cerebral palsy

Most children with CP are born with a normal spine. However, their spine may be exposed to abnormal forces which when left unmanaged may lead to lateral deviation from the midline, postural asymmetry, and the development of neuromuscular scoliosis (51). Reported prevalence of neuromuscular scoliosis in children with CP varies between 11% and 64% (52-54) depending on how it has been defined, the severity of CP and their age.

Neuromuscular scoliosis most often presents with a 'C-shaped' curvature (55, 56), commonly in the thoracolumbar region (57), and will continue to progress in adulthood, beyond the skeletal maturity of the individual (51, 58). Scoliosis can result in difficulties with sitting, balance (59, 60), breathing (61, 62), swallowing (55), digestion, skin integrity (63, 64) and pain (54, 64). Risk factors for developing scoliosis include higher GMFCS level (62, 65), female sex, epilepsy, limited hip or knee flexion (66), and spasticity (7, 63, 67).

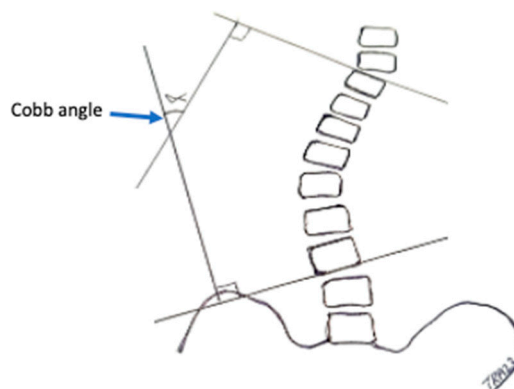


Figure 6. Measuring Cobb angle

Scoliosis is detected at a radiographic examination with Cobb angles $\geq 10^{\circ}$ (55, 56, 68), or clinically using the Adams Forward Bend test. The Cobb angle is the most common measurement used to determine the severity of scoliosis; and is typically used to decide both the progression and the need for orthopaedic treatment (69). The Cobb angle is calculated by drawing lines across the most tilted vertebrae endplates above and below the apex of the curve (70), then perpendicular lines drawn from each of these first lines, and the angle taken where they intersect, as shown in the diagram on the previous page (Figure 6).

In this thesis in Studies III-IV, classification of scoliosis includes having Cobb angles of $\geq 20^{\circ}$ or having a moderate or severe curve on clinical examination. The use of clinical examination has been shown to have high inter-rater reliability (kappa 0.96), sensitivity of 75%, and specificity of 95.8% when compared to radiographic Cobb angles of $\geq 20^{\circ}$ (71), and subsequently only those children with moderate and severe curves on clinical examination are referred for further radiographic examination.

Biomechanical mechanism of scoliosis

The scoliosis evolves from a flexible curve to a fixed spinal deformity during the child's growth period and continues to progress even after skeletal growth has finished (72). The biomechanics of scoliosis development and progression can be explained in part by the Hueter-Volkman principle. Development of scoliosis progresses from a lateral curve to become a 3-dimensional spinal deformity with rotation (59, 73). This principle identifies how mechanical compression on one side of the epiphyseal growth plate retards bony growth, whilst traction on the other side can stimulate growth. This together with an initial curvature or an asymmetric loading or compression results in asymmetric growth. This leads to an imbalance of growth across the vertebrae, resulting in vertebral wedging, and a further asymmetric posture, loading and spinal adaptation. This pattern will generally continue if left unmanaged and is often referred to as a 'vicious cycle' of progressive deformity of the spine (73-75) (Figure 7).

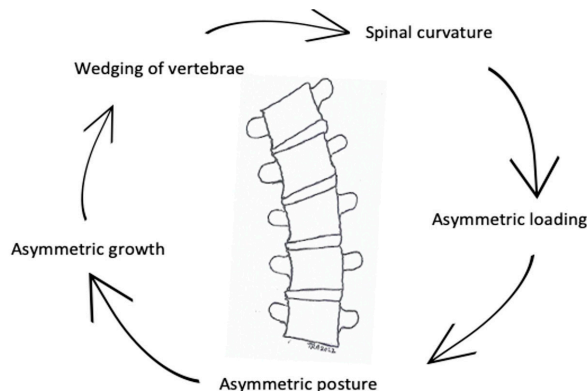


Figure 7. Illustration showing Hueter-Volkman's principle in action on the vertebral column.

Windswept hip deformity

Hip disorders can be common amongst children with CP, despite the hips typically presenting as normal at birth (76, 77). If left unmanaged ROM limitations, hip subluxation or even dislocation may develop causing pain or negatively impact upon the ability to sit, lie, stand or walk, or completion of personal care activities. One such disorder now commonly referred to as windswept hip deformity (WSH) is a clinical manifestation exhibited in some children. It was first described by Fulford and Brown (40) as windswept child syndrome, and then as windblown hip syndrome by Letts et al. (78) and presents with one hip abducted and externally rotated whilst the other hip is adducted and internally rotated.

It has been suggested that WSH may follow hip subluxation or dislocation (8, 78), or it may be preceded by the occurrence of scoliosis, resulting in asymmetric postures, discomfort, and restricted functional participation. Although, equally, the presence of WSH may precede the development of scoliosis (62). However, in Casey et al. (54) there were so few hip dislocations in the sample, perhaps it may be possible that knee contracture could also be considered a trigger for WSH (41, 79). Additionally, a low prevalence of hip dislocations has been attributed to the success of the national hip surveillance program (80), with early monitoring through clinical and radiographic examinations. WSH can be difficult to treat (8), and hence the need to regularly monitor the hips and try to prevent it from becoming established.

In studies III-IV we included children with all types of CP, and not just those with spastic bilateral CP. The Persson-Bunke method of identifying WSH was used to determine presence of WSH. In order to have WSH children had $\geq 50\%$ difference either in the abduction, internal, or external rotation ROM between left and right hips (8). This method offers an objective measurement of WSH based upon the hip ROM, as opposed to the postural presentation of the child which could be influenced by positioning and a lack of ability to move. WSH prevalence has been reported as 9% (54, 79). With direction of WSH being classified using this method, the picture below illustrates what was calculated to be a right WSH with femurs and knees pointing towards the right side of the child (Figure 8).

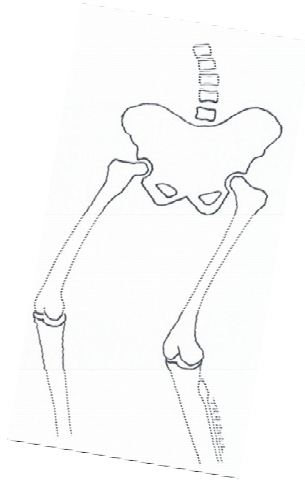


Figure 8: Illustration frontal view of a right windswept hip deformity

Contractures of the hips and knees

Contractures are frequently observed in those with an upper motor neuron lesion, such as CP (3, 81), and previously were often reported to be a result of spasticity (82, 83). However, the mechanics behind the development of contractures are recognised as complex (84), and that there is more than spasticity at play as some individuals without spasticity will present with contractures (3). It is now understood that in addition to spasticity, that other variables contributing to contracture development include muscle weakness, immobility, and muscle pathology (85, 86).

Although clinicians give much attention to the prevention of contractures, children with CP continue to develop them in both upper and lower limbs (54, 85, 87). Once developed they restrict not only the ROM, but also the ability of the child to grasp with their hands, lie, stand, and walk comfortably. They can also be painful (81, 85) to the individual; and can make completion of toileting and washing activities difficult (88).

Contractures occur when there is loss of tissue elasticity (3), and a shortening of these tissues (89) over a prolonged period and with the presence of lengthened sarcomeres and fewer satellite cells (85, 86), resulting in a loss of ROM around a joint (89) and the contracture developing.

Link between postural asymmetries, deformities, and contractures

Early work by Fulford and Brown (40) investigating ‘squint’ baby syndrome and whether the deformities seen diminish over time suggested that there was an association between positioning of the infant, the influences of gravity and immobility (49), and the development of deformities. They also suggested that the position of one body segment can have an impact on the position of another body segment and can lead to positional deformity (49), or what we also refer to as postural asymmetry of the child (90). Later work by Letts et al. (78) concurs with the possibility that alignment of one body segment impacts upon another in their work exploring temporal relationship of scoliosis, pelvic obliquity and hip dislocation. Further, Porter et al. (90) also report that there is a relationship between the position in lying in the early years of the child and the development of postural asymmetries and deformities.

Therapeutic management

Clinical therapeutic intervention aims to use the principles of 24-hour postural management with these children to promote the structural integrity and functioning of the musculoskeletal system, prevent postural asymmetries, joint contractures and deformities occurring or progressing (91, 92). Clinicians frequently focus on positioning in sitting (93) and lying (94, 95) where many children are unable to self-correct their position effectively or even at all. Although interventions may also include the use of orthotic devices (72, 96, 97) and standing programmes (98). It is vital that clinical and postural interventions commence early for these children (99) to minimize or delay the need for surgical interventions (100).

Chronic hip subluxation can result in marked hip damage, including dislocation, and pain for children (101-104), and if left to progress then often in adulthood there is degenerative arthritis (105) and a need for hip salvage surgery. Scoliosis curves can progress can cause respiratory compromise, gastrointestinal difficulties, pain, reduced sitting balance, reduced quality of life and even premature death (52, 55, 62, 97, 106, 107). These result in reduced functioning, participation in everyday activities (101), and quality of life, and hence why it is so important that we attempt to prevent this from occurring for persons with CP.

Registry-based studies

There are many benefits as well as limitations to registry-based studies. This doctoral project has been able to access data of a total population of children with CP living in Sweden. Registry based studies provide access to a large, unselected dataset, reducing the risk of selection bias in recruitment and sampling (108), and allowing meaningful ‘real-World’ data analysis. Data is collected prospectively in a systematic manner, and with repeated measurements this allows analysis of trends and patterns of both treatments and outcomes. Data can be analysed using a variety of methodologies, including cross-sectional, longitudinal.

Further, registry-based studies allow for greater generalisation and comparison of results between countries with similar health and social care systems, or similar populations being studied. This doctoral project enables a comprehensive overview of the whole population of children with CP, and across all GMFCS levels. The use of registry-based studies can provide a comprehensive picture of intervention delivered by clinicians as a whole working with these children, rather than focusing on a particular clinic or service.

In antithesis, registries tend to lack some details (for example, the specific make, model of wheelchair and type of postural seating within it is not available), and so the research questions are limited to what data is available.

There may be some missing assessment data. For example, perhaps a child may have become unwell or non-cooperative with the therapist during completion of an assessment and it could not be fully completed or had to be discontinued; or occasionally there could have been a loss of data due to registry system failure. However, the CPUP national registry and follow-up program has excellent engagement by therapists and families alike, with >95% of all children in Sweden with CP born 2000 onwards, being included in the annual/ biannual assessments (109)(Figure 9).

In efforts to mitigate against possible measurement or recording errors, clinicians receive local training in how to complete the assessments, and written manuals and guidelines of how to measure and report results into the database are available. Further, the registry team complete an annual cleaning of the data, validating much of the data against results from more than one outcome measure.

CPUP: The follow up program for cerebral palsy

The CPUP is a national registry and follow-up programme of children with cerebral palsy in Sweden which commenced initially in Southern Sweden in 1994. It was later classified as a National Healthcare Quality Registry in 2005 covering all of Sweden, and has >95% of all children with CP born 2000 to present reported into this database (110). The primary goal of the programme is the prevention of both hip dislocation and severe contractures (100) in children with CP. An additional

aim of the programme was to describe the progression and functioning of individuals with CP over time, as well as to facilitate increased inter-professional working (110).

Children with, or with suspected CP are included in the registry as early as possible, with diagnosis being confirmed by neuropaediatricians from the child's 4th birthday according to the inclusion/ exclusion criteria of SCPE (14). If the child does not meet the inclusion criteria, they are then excluded from the registry (87, 109).

The children are reviewed and assessed annually or biannually depending upon their age and GMFCS level, as shown in Figure 9, by their local physiotherapists and occupational therapists. Data from all other healthcare professionals involved in the child's care is also recorded in the database.

As a result of this national surveillance programme the number of hip dislocations (80), scoliosis (62) and severe contractures (100) has been shown to decrease since its introduction. Subsequently programmes using similar assessments as in CPUP have been established in other countries, including, Norway (2006), Denmark (2010), Iceland (2012), New South Wales in Australia (2012), Scotland (2013) (111). Whilst other CP registries exist, such as that in Northern Ireland (1991)(112), they do not gather the same level of clinical detail as contained within the CPUP. The CPUP evolved to include adults in 2009.

The GMFCS was introduced in 1997 and was then universally accepted as a classification of gross motor performance and utilised within CPUP from this date. The PPAS was included with all children from 2017 onwards. Therefore, there was no data on postural presentation as described using the PPAS to allow longitudinal follow-up prior to 2017.

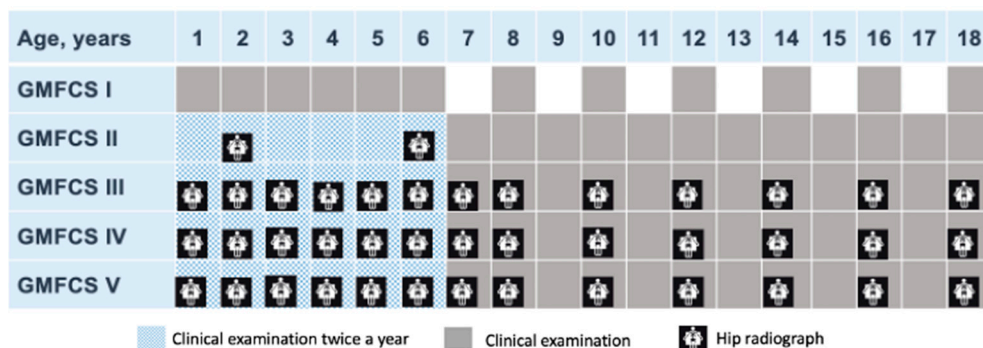


Figure 9. Guidelines for clinical examination and hip radiograph within the Swedish Cerebral Palsy Follow-up Program (CPUP). This figure is reprinted with permission from Dr Roddy-Bousquet.

Aims

The aims of this thesis were to provide further information about postural asymmetries, deformities, contractures, and pain for children with cerebral palsy in sitting, in supine and when using a wheelchair so that suitable interventions can be offered to improve and enable their activity and participation in everyday life.

- Study I* To analyse which physical factors influence independent use of manual and power wheelchairs.
- Study II* To examine for associations between postural asymmetries, postural ability, and pain in supine and sitting positions.
- Study III* To determine the prevalence of deformities and contractures of the spine, hips, and knees, and their association with postural asymmetries, ability to change position, and pain, considering differences in sex, age, and gross motor function.
- Study IV* To analyse which comes first, scoliosis or windswept hip deformity.

Materials and methods

Study design

Studies I-III were cross-sectional studies of children with CP from the Swedish National Follow-up programme and registry for individuals with CP (CPUP). *Study I* used a total population, whilst *Studies II-III* excluded those with missing data of the primary outcomes posture or ROM, respectively. *Study I* described how many children aged 0-11 years use manual and power wheelchair mobility indoors and outdoors, as well as the risk factors for not being able to do so independently. *Studies II-III* with children aged 0-18 years looked at the relationship between postural asymmetries, postural ability, and pain (*study II*), and the prevalence and association of scoliosis, windswept hips, knee and hip flexion contractures (*study III*). *Study IV* was a longitudinal cohort study identifying if scoliosis or windswept hip deformity occurred first for children born 1990-2018.

Participants and methods

Study I

Data for all 2,328 children with CP aged 0-11 years (58% boys and 42% girls) born 2002-2013 and reported into the national CPUP registry was included. The latest occupational therapy and physiotherapy examinations for each child performed between January 1st, 2012, and June 30th, 2014, was extracted and used for analysis.

The use of manual and power wheelchair indoors and outdoors was analysed in relation to age, sex, GMFCS and MACS levels; and the influence of upper extremity ROM, thumb-in-palm deformity, wrist and finger flexor spasticity (Zancolli), House functional classification system, and bimanual ability on wheelchair use examined.

Study II

Included 2,735 children with CP aged 0-18 years (59.5% boys and 40.5% girls) reported into the CPUP between January 1st, 2017, and June 30th, 2018, with data for posture in sitting and supine lying. The Posture and Postural Ability Scale data was used to identify postural asymmetries and their ability to maintain and change position in sitting and supine. Postural asymmetry was grouped into four

categories: having “severe” postural asymmetry (0-1 points), “moderate” asymmetries (2-3 points), “mild” asymmetries (4-5 points), or “full” symmetry (6 points). Similarly postural ability was grouped into four categories: children who had severe postural deficit as “unable to maintain position” (PPAS levels 1-2), those with moderate postural deficit as “maintains position” (PPAS levels 3-4), those with mild postural deficit as “moves within position” (PPAS levels 5-6), and those children able to independently move in and out of sitting or supine as “changes position” (PPAS level 7). Postural ability in sitting and supine was also dichotomised into “unable to change position” (PPAS levels 1-6) and “able to change position” (PPAS level 7). The presence of pain was self-reported as a “yes/no” by either the child or their parent/ caregiver; and age was categorised into six groups (0-3, 4-6, 7-9, 10-12, 13-15, 16-18 years).

Study III

This study included 2,450 children aged 0-18 years from the same CPUP reports as Study II, with the addition of complete data for scoliosis, hip and knee ROM. Included were 990 girls (40.4%) and 1,460 boys (59.6%) with a mean (SD) age of 9.4 (4.3) years.

As part of the Follow-up program the children’s local physiotherapists completed the assessments of the spine, hips, and knees. Scoliosis was measured on clinical examination using the Adam’s forward bend test. Scoliosis was defined as moderate or severe curve, or as having had a spinal fusion.

Passive range of motion (ROM) of the hips and knees was completed using standardised goniometric procedures (www.cpunp.se). Specifically, abduction, adduction, external and internal rotation, flexion and extension was measured for each hip, along with ROM for knee flexion and extension (54). A loss of passive hip or knee extension of $\geq 10^{\circ}$ ROM was used to classify a hip or knee flexion contracture, whilst an inability to flex the hips to 90° was classified as a hip flexion limitation. This cut-off value was used in Studies III-V as this (1) is a clinically significant loss of ROM that can have a functional consequence for the child; and (2) is visually observable, reducing potential impact of measurement error by the clinician. Inability to flex either knee to 90° was classified as a knee contracture. Windswept hip deformity was calculated using the Persson-Bunke method (71) based on the passive ROM of abduction, internal and external rotation of the hips, with a difference of at least 50% between the left and right side.

Study IV

This study was a longitudinal cohort study based on 41,600 measurements for 4,148 children born January 1st, 1990, to December 31st, 2018, and enrolled in the national registry and follow-up program before 6 years of age. Children were followed prospectively for up to 26 years.

Having scoliosis was confirmed through clinical examination defining it as moderate or severe, radiographic Cobb angles of $\geq 20^{\circ}$, or having had spinal fusion.

Windswept hip deformity was calculated based on passive ROM of the hips based on the Persson-Bunke's method (113). Receiving intrathecal baclofen pump, selective dorsal rhizotomy, pelvic and hip surgeries prior to the onset of scoliosis or windswept hip deformity were used to censor the child's follow-up in the study as these may have influenced the occurrence of scoliosis or windswept hip deformity.

Statistics

For *Study I* all analyses were performed using STATA 12 software; for *Studies II-III* IBM SPSS Statistics 26 was used. For *Study IV* SPSS Statistics 26 was used to collate and merge the data, and R ≥ 4.0 (R Foundation for Statistical Computing, Vienna, Austria) for analyses. P-values < 0.05 were considered significant for all statistical analyses in this thesis. Since the underlying population distribution may not be considered approximately normally distributed, non-parametric statistical tests were used in our analyses.

In *Study I-IV*, for descriptive statistics, categorical data were reported using frequencies (n) and percentages (%), whilst continuous data were reported as means with accompanying standard deviations (SDs).

In *Study I*, Cox Proportional Hazard model with an equal follow-up and robust variance were used. The resulting hazard ratios (HRs) with 95% Confidence Intervals (CIs) and p-values were used to determine the risk for not being able to self-propel, for each of the analysed risk factors. The estimated influence of the analysed risk factor on the outcome in the model was adjusted for sex and age. HRs are used to specify the explanatory variable's effect on the risk of an event occurring.

In *Study II*, for categorical variables Pearson's χ^2 -test and chi-square (χ^2) test for trend were used for tests of differences between the categorical variables, whilst Spearman's rank correlation r_s was used to estimate correlations.

In *Study II-III*, simple and multiple logistic regression analyses were used to estimate the magnitude of associations between the predictor variables and the outcome variable, with the results presented as odds ratios (ORs) with accompanying 95% CIs. In *Study II*, logistic regression analysis was used to estimate the magnitude of associations between postural asymmetries and pain. In *Study III*, logistic regression analysis was used to estimate the magnitude of associations between sitting and lying posture, ability to change position, pain, range of motion of lower limbs, and deformities. ORs are used to estimate the risk of observing a particular outcome when there are at least two possible outcomes.

In *Study IV*, one-sample χ^2 -test was used to examine if the distribution of children among the GMFCS levels and CP subtypes within each event group differed from what could be expected based on the overall distribution of children over GMFCS levels and CP subtypes. Kruskal-Wallis test was used to examine differences in age

at first event between GMFCS levels and CP subtypes, respectively, within each event group.

Ethics

Ethical approval was granted by the Medical Research Ethics Committee at Lund University for all four studies (Dnr 443-99, 383/2007, 2011/9), and permission to extract data from the registry was obtained by the Samrådsgrupp för Kvalitetsregister, vårdinformationssystem och beredning (KVB) Region Skåne and the registry holder at:

<https://vardgivare.skane.se/kompetens-utveckling/forskning-inom-region-skane/utlamnande-av-patientdata-samradkvb/>.

Results

Study I: Physical risk factors influencing wheeled mobility

A total of 2,328 children aged 0-11 years were reported from January 2012 to June 2014. Of these 1,344 (58%) were boys, and the mean (SD) age was 6 years 2 months (2 years, 11 months). No child used a manual wheelchair before 1 year, and none used a power wheelchair before 3 years of age.

Wheelchairs indoors were used by 610 children (26%), and of these 537 (88%) used only manual, 11 (2%) used only power, and 62 (10%) used both manual and power wheelchairs indoors. Of the manual wheelchair users, only 28% were able to independently self-propel indoors; and 66% were able to independently drive their power chair indoors. For outdoors use, only 10% were able to self-propel their manual, whilst 75% were able to drive their power chair independently (Table 6).

Table 6. Distribution of wheelchair use, note that some children use both manual and power

Wheelchair	INDOORS (n=610)		OUTDOORS (n=858)	
	n	%	n	%
MANUAL	(599)		(838)	
Self-propels	165	28	83	10
Pushed	434	72	755	90
POWER	(71)		(166)	
Drives independently	47	66	125	75
Needs assistance	24	37	41	25

Risk factors for not being able to independently self-propel a manual wheelchair indoors were having poor hand function (HR 2.4-5.4), finger and wrist extensor spasticity (HR 2.2) and high GMFCS levels (HR 2.6-4). As levels of function decreased the risk for not self-propelling increased. Shoulder flexion and wrist extension limitations had greater impact on manual wheelchair use indoors than having limited supination or elbow extension (Table 7).

The greatest risk factor for not being able to independently drive a power wheelchair indoors was being classified as GMFCS level V (HR 12.6) or MACS IV-V (HR 9.8). The risk for not driving a powered wheelchair independently outdoors was much greater for children with reduced hand function classified as House 0-3 (HR 20.7), finger and wrist flexors spasticity (HR 5.4), limited shoulder and wrist ROM (HR 5.5-6.6) and being classified as GMFCS level V (HR 25.8) (Table 7).

Table 7. Hazard ratios (HR) with 95% confidence intervals (CI) and p-values (P) adjusted for age and sex. Variables presented are treated as risk factors for not being able to self-propel

Physical factors	Manual wheelchair indoors			Power wheelchair indoors			Manual wheelchair outdoors			Power wheelchair outdoors						
	HR	95% CI	P	HR	95% CI	P	N	HR	95% CI	P	N	HR	95% CI	P	N	
GMFCS IV	2.6	1.8-3.7	<0.001	590	1.8	0.3-12	0.522	75	1.1	1-1.1	0.082	829	5.7	1.4-24	0.017	168
GMFCS V	4	2.9-5.6	<0.001		13	2.2-70	0.004		1.2	1.1-1.3	<0.001		26	6.5-103	<0.001	
MACS III	2.4	1.5-3.9	0.001	545	0.6	0.04-8.2	0.673	74	1.2	1.1-1.4	<0.001	768				164
MACS IV-V	5	3.3-7.8	<0.001		9.8	1.6-61	0.015		1.4	1.3-1.5	<0.001					
House 4-6	3.3	2-5.6	<0.001	546	1.1	0.2-8.4	0.894	73	1.3	1.1-1.4	<0.001	767	5.9	0.8-43	0.083	163
House 0-3	5.4	3.3-8.9	<0.001		7.3	1.2-42	0.028		1.4	1.2-1.5	<0.001		21	3-145	0.002	
Zancollì 2-3	2.2	1.8-2.5	<0.001	540	5.4	1.9-15	0.001	71	1.2	1.1-1.3	<0.001	762	5.4	2.5-11	<0.001	159
Thumb-in palm I-IV	1.6	1.4-1.8	<0.001	539	1.6	0.7-4	0.271	70	1.1	1.1-1.2	<0.001	757	1.6	0.9-2.9	0.116	161
Bimanual ability	0.5	0.4-0.5	<0.001	558	0.2	0.1-0.4	<0.001	74	0.9	0.8-0.9	<0.001	783	0.2	0.1-0.3	<0.001	164
Shoulder flex >120 to <160	1.1	1-1.3	0.059	554	1.8	1-3.5	0.065	74	1.1	1-1.1	0.094	775	2.1	1.2-3.9	0.014	164
Shoulder flex <=120	1.4	1.3-1.5	<0.001		3.6	2.2-5.8	<0.001		1.1	1.1-1.2	<0.001		5.5	3.4-8.7	<0.001	
Elbow ext >-30 to <-10	1.2	1-1.4	0.019	558	1.8	0.7-4.5	0.185	74	1	0.9-1.1	0.762	778	2.9	1.4-5.9	0.003	162
Elbow ext <=-30	1.3	1.1-1.5	0.005		2.9	1.7-4.8	<0.001		1	0.9-1.2	0.519		4.1	2.4-7	<0.001	
Supination >45 to <80	1.2	1-1.3	0.029	556	0.8	0.3-2.7	0.777	74	1.1	1-1.1	0.068	779	1.8	0.9-3.7	0.088	164
Supination <45	1.1	0.9-1.3	0.232		2	1-4.1	0.047		1.1	1-1.2	0.024		2.1	1-4.4	0.045	
Wrist ext >0 to <60	1.1	1-1.3	0.045	552	2.4	1.1-5.4	0.030	74	1	0.9-1	0.809	772	2	1-4	0.050	161
Wrist ext <=0	1.5	1.3-1.7	<0.001		5	2.2-11	<0.001		1.1	1-1.2	0.117		6.6	3.2-14	<0.001	

Note: GMFCS I-III, MACS I-III, House 7-8, Zancollì 1, no thumb-in-palm, no bimanual ability and "green" values for range of motion were used as reference categories. Statistically significant (p<0.05) Hazard ratios (HR) of 1-1.2 indicating an increased risk of 10-100% are marked with yellow, HRs of >2 (more than doubled risk) are marked with red and HRs <1 (meaning a reduced risk is indicated with green)

Study II: Postural asymmetries, pain, postural ability

There was data on postural asymmetries and postural ability and in supine and sitting for 2,735 of the 3,296 children aged 0-18 years, with 1,628 boys (59.5%) and a mean (SD) age of 9.2 (4.4) years. Over half of the 2,735 children had postural asymmetries, with more present in sitting ($n = 1,646$; 60.2%), than in supine ($n = 1,467$; 53.7%).

Postural asymmetries were seen across all GMFCS levels. Children at GMFCS levels I-II more frequently had either symmetric or mild postural asymmetries (involving one or two body segments) in supine and sitting positions, whilst those at GMFCS level III had moderate asymmetries, and those with GMFCS levels IV-V had severe postural asymmetries (with total body involvement). For children with GMFCS levels I-IV the most common location was the upper and lower extremities in supine, and for children with GMFCS level V it was the head and trunk. In contrast for sitting, the most common postural asymmetries involved the trunk, pelvis, and weight distribution for those children at GMFCS levels I-IV, and for the GMFCS level V the whole body. A total of 824 (30.1%) children were unable to independently move in and out of a supine position, whilst 995 (36.4%) were unable to independently move in and out of a sitting position. Most children who were unable to independently change position had an asymmetric posture in supine and sitting.

Children who could move but not change position in supine were more than twice as likely (OR 2.62, CI 1.8-3.82) to have postural asymmetries, whilst those in sitting were 1.5 times more likely (Table 8), even when adjusted for all other variables in the model. As postural ability skills decreased in sitting or supine positions the children were 4 to 7 times more likely to have postural asymmetries (OR 4.15-7.79). The risk of having postural asymmetries increased with higher GMFCS levels (Table 8).

Pain prevalence was reported by 1,036 / 2,640 (39.2%) of the children. Having severe postural asymmetries in sitting and supine sagittal views doubled the risk for pain (Table 9). Children also had a higher OR for pain when they were unable to change position (OR 1.5 to 2.3) and with increased age (OR 1.08; 95% CI 1.06-1.10).

Table 8. Logistic regression analyses with odds ratios (ORs) and 95% confidence intervals (CI) for asymmetric postures (PPAS 0-5 points) in sitting and supine positions

	Asymmetric supine posture				Asymmetric sitting posture			
	OR	95% CI		P-value	OR	95% CI		P-value
Changes position (7)	Ref.				Ref.			
Moves within position (5-6)	2.62	1.8	3.82	<0.001	1.5	1.02	2.21	0.04
Maintains position (3-4)	3.45	1.9	6.27	<0.001	3.22	1.87	5.55	<0.001
Cannot maintain position (1-2)	7.79	2.19	27.73	0.002	4.15	2.3	7.48	<0.001
Female sex	1.24	1.01	1.52	0.036	1.14	0.94	1.38	0.178
Age	1.07	1.05	1.1	<0.001	1.05	1.03	1.07	<0.001
GMFCS I	Ref.				Ref.			
GMFCS II	2.79	2.15	3.62	<0.001	2.42	1.9	3.08	<0.001
GMFCS III	5.48	4.09	7.35	<0.001	3.32	2.42	4.54	<0.001
GMFCS IV	11.62	8	16.9	<0.001	7.17	4.53	11.36	<0.001
GMFCS V	27.82	14.49	53.43	<0.001	19.34	9.22	40.59	<0.001

Note: PPAS, Posture and Postural Ability Scale. Ability to move into and out of position independently in supine or sitting, Gross Motor Function Classification System (GMFCS) level I and being male were used as reference categories. Age was used as a continuous variable. All variables were adjusted for all other variables in the model.

Table 9. Simple logistic regression analysis with odds ratios (ORs) and 95% confidence intervals for pain in children with mild, moderate and severe postural asymmetries in supine and sitting, frontal and sagittal views.

POSTURAL ASYMMETRIES	SUPINE OR (95%CI)		SITTING OR (95%CI)	
	Frontal	Sagittal	Frontal	Sagittal
Mild	1.05 (0.8-1.3)	1.16 (0.9-1.4)	1.33 (1.1-1.7)	1.27 (1.0-1.6)
Moderate	1.48 (1.1-2.0)	1.33 (1.0-1.8)	1.41 (1.1-1.9)	1.73 (1.3-2.2)
Severe	1.86 (1.3-2.7)	2.12 (1.5-3.1)	1.88 (1.3-2.7)	1.97 (1.3-3.0)

Note: adjusted for age, sex and GMFCS levels

Study III: Deformities and contractures

Of the 2,450 children included with ROM and posture data 1,460 (59.6%) were boys, with a mean (SD) age of 9.4 (4.3) years. Scoliosis was seen in 257 children (10.5%), windswept hip deformity (WSH) in 213 (8.7%), knee flexion contractures of $\geq 10^\circ$ in 470 (19.2%), hip flexion contractures in 162 (6.6%), less than 90° of hip flexion in 25 (1%) and dislocated hips in 5 children (0.2%). These latter two sub-groups with too few observations were omitted from further analyses.

Deformities and contractures of the spine, hip and knee were seen across both sexes, all age groups and all GMFCS levels. Although the likelihood of having scoliosis was higher in girls (OR 1.7, CI 1.2-2.3) and increased with GMFCS-level and age (Table 10).

Children with postural asymmetries in supine or sitting had a higher risk for/likelihood of scoliosis, WSH, hip and knee flexion contractures even when adjusted for all sex, age-group, and GMFCS level. Having an asymmetric supine posture increased the likelihood of having windswept hips (OR 8.8, CI 4.9-15.8), hip flexion contractures (OR 6.7, CI 3.4-13.1) and knee flexion contractures (OR 12.2, CI 7.1-20.8); whilst having an asymmetric sitting posture increased the likelihood of having scoliosis (OR 9.1, CI 4.9-16.9) and windswept hips (OR 5.7, CI 3.2-9.8), when adjusted for age, sex, and GMFCS level. Further, having windswept hip deformity (OR 1.6, CI 1.2-2.2) and hip flexion contractures (OR 1.5, CI 1.1-2.1) were significantly associated with having pain when adjusted for age, sex and GMFCS level (Table 10).

Table 10. Logistic regression analysis with adjusted odds ratios (ORs) and 95% confidence intervals (CIs) for deformities and contractures

	Scoliosis (Sitting Frontal)			Windswept hips (Sitting Frontal)			Windswept hips (Supine Frontal)			Hip Flexion Contracture (Supine Sagittal)			Knee Flexion Contracture (Supine Sagittal)							
	OR	95% CI	p-value	OR	95% CI	p-value	OR	95% CI	p-value	OR	95% CI	p-value	OR	95% CI	p-value					
Sex																				
Boy	<i>Ref.</i>			<i>Ref.</i>			<i>Ref.</i>			<i>Ref.</i>			<i>Ref.</i>							
Girl	1.7	1	2.3	0.001	1.2	1	1.7	0.17	1.3	1	1.7	0.12	1	1	1.4	0.93	0.8	1	1.1	0.15
Age group																				
0-3 yrs	<i>Ref.</i>			<i>Ref.</i>			<i>Ref.</i>			<i>Ref.</i>			<i>Ref.</i>			<i>Ref.</i>			<i>Ref.</i>	
4-6 yrs	2.2	1	5.4	0.09	2.2	1	5.4	0.11	2.2	1	5.7	0.09	4	1	18	0.07	3.2	2	6.7	0.001
7-9 yrs	5	2	12	<0.001	2.8	1	7	0.03	2.8	1	6.9	0.03	6.6	2	29	0.01	5.2	3	11	<0.001
10-12 yrs	6.9	3	16	<0.001	4.5	2	11	0.001	4.3	2	10	0.001	9.7	2	41	0.002	11	5	21	<0.001
13-15 yrs	11	5	25	<0.001	6.8	3	16	<0.001	6.1	3	15	<0.001	16	4	66	<0.001	20	10	40	<0.001
16-18 yrs	14	6	33	<0.001	8.7	4	22	<0.001	8	3	20	<0.001	18	4	78	<0.001	19	9	41	<0.001
GMFCS level																				
GMFCS I	<i>Ref.</i>			<i>Ref.</i>			<i>Ref.</i>			<i>Ref.</i>			<i>Ref.</i>			<i>Ref.</i>			<i>Ref.</i>	
GMFCS II	1.5	1	3.3	0.31	1.1	1	2	0.66	1.1	1	1.9	0.8	1.6	1	3.2	0.14	1.8	1	3.3	0.07
GMFCS III	3.1	2	6.4	0.002	1.8	1	3.2	0.03	1.6	1	2.8	0.1	2	1	4	0.06	8.5	5	15	<0.001
GMFCS IV	5.5	3	11	<0.001	1.2	1	2	0.56	0.9	1	1.6	0.78	2.2	1	4.2	0.02	15	9	25	<0.001
GMFCS V	16	9	32	<0.001	2.8	2	4.9	<0.001	2	1	3.5	0.02	2.9	2	5.7	0.002	14	8	24	<0.001
Asymmetry																				
No (6 p)	<i>Ref.</i>			<i>Ref.</i>			<i>Ref.</i>			<i>Ref.</i>			<i>Ref.</i>			<i>Ref.</i>			<i>Ref.</i>	
Mild (4-5 p)	3.1	2	5.6	<0.001	1.6	1	2.6	0.05	2	1	3.3	0.004	2.2	1	3.9	0.007	4.7	3	7.2	<0.001
Moderate (2-3 p)	5	3	8.9	<0.001	3.4	2	5.6	<0.001	4.8	3	8.2	<0.001	4.9	3	9.1	<0.001	11	7	17	<0.001
Severe (0-1 p)	9.1	5	17	<0.001	5.7	3	9.8	<0.001	8.8	5	16	<0.001	6.7	3	13	<0.001	12	7	21	<0.001

Note: Posture and Postural Ability Scale (PPAS) no asymmetry in sitting or supine, Gross Motor Function Classification System (GMFCS) level 1, youngest age group (0-3 years), and being a boy were used as reference categories. Age was used as a categorical variable. All variables were adjusted for all other variables in the model.

Study IV: Which develops first: scoliosis or windswept hip deformity?

For the 4,148 children born 1990 to 2018, reported into the registry before they were six years old there were a total of 41,600 measurements over a period of 26 years. These children had a mean (SD) age at first examination of 2.8 [1.4] years and 58.3% were boys. Most children were classified at GMFCS level I (n = 1,912; 46.1%), and there was a higher proportion with spasticity (n = 3,299; 79.5%) (Table 11). Most children did not develop either scoliosis or WSH during the follow-up period (n = 2,577, 62.1%).

Table 11. Distribution of CP Sub-type by GMFCS level at 5 years of age

CP subtype	GMFCS level										Total n
	I		II		III		IV		V		
	n	%	n	%	n	%	n	%	n	%	
Ataxia	73	4.1	46	25.8	40	22.5	15	8.4	4	2.2	178
Dyskinesia	27	5.6	40	8.3	35	7.3	153	31.9	225	46.9	480
Spasticity	1753	53.1	472	14.3	293	8.9	392	11.9	389	11.8	3299
UC/ Mixed	59	30.9	20	10.5	17	8.9	35	18.3	60	31.4	191
Total	1912	46.1	578	13.9	385	9.3	595	14.3	678	16.3	4148

Note: Unclassified (UC); Gross Motor Function Classification System (GMFCS)

Overall, there was a greater incidence of WSH (16.6%) than scoliosis (8.1%), with more children having WSH as a first deformity. Children with GMFCS levels I-II were more likely to develop WSH first. Whilst for children with GMFCS level V, the likelihood of developing either deformity (scoliosis 19.8%, WSH 21.5%) was spread more evenly. The children with GMFCS level V were also more likely to have surgery first (31.7%) before developing either deformity, and also to have surgery at a younger age (mean [SD], 4.8 [2.3] years) compared to the other GMFCS levels (Table 12).

We found that children who had dyskinesia were the most likely to develop either of the deformities, whilst those least likely were those with ataxia (Figure 10). However, when children with ataxia did develop a deformity there was a higher probability of developing scoliosis (9%) when compared to WSH (7.3%). Those with spasticity and dyskinesia were more likely to develop WSH (17% and 20% respectively). However, children with dyskinesia had a similar incidence of developing WSH (20%) as scoliosis (18%).

Table 12. First event and age at first event according to Gross Motor Function Classification System (GMFCS) level at 5 years of age for the n = 4,148 participants.

Variable	GMFCS level				
	I (n = 1912)	II (n = 578)	III (n = 385)	IV (n = 595)	V (n = 678)
First event, n (%)					
- Scoliosis	55 (2.9)	30 (5.2)	34 (8.8)	83 (13.9)	134 (19.8)
- Windswept hips	240 (12.6)	110 (19.0)	74 (19.2)	117 (19.7)	146 (21.5)
- Tied scoliosis/windswept hips	3 (0.2)	1 (0.2)	0 (0.0)	9 (1.5)	23 (3.4)
- No event during follow-up	1582 (82.7)	403 (69.7)	199 (51.7)	233 (39.2)	160 (23.6)
- Censored at surgery	32 (1.7)	34 (5.9)	78 (20.3)	153 (25.7)	215 (31.7)
Age at first event, mean (SD)					
- Scoliosis	12.0 (5.8)	9.9 (4.7)	11.0 (4.1)	8.0 (4.0)	5.5 (3.5)
- Windswept hips	9.4 (4.8)	8.7 (5.4)	10.2 (5.6)	7.2 (4.5)	5.2 (3.3)
- Tied scoliosis/windswept hips	9.8 (3.1)	3.4 (-)	- (-)	8.6 (4.4)	5.6 (3.0)
- No event during follow-up	9.6 (5.0)	9.1 (5.1)	8.3 (4.8)	8.0 (5.1)	5.5 (3.8)
- Censored at surgery	8.4 (3.7)	7.1 (3.5)	5.4 (2.7)	5.7 (2.8)	4.8 (2.3)

Note: SD, Standard deviation

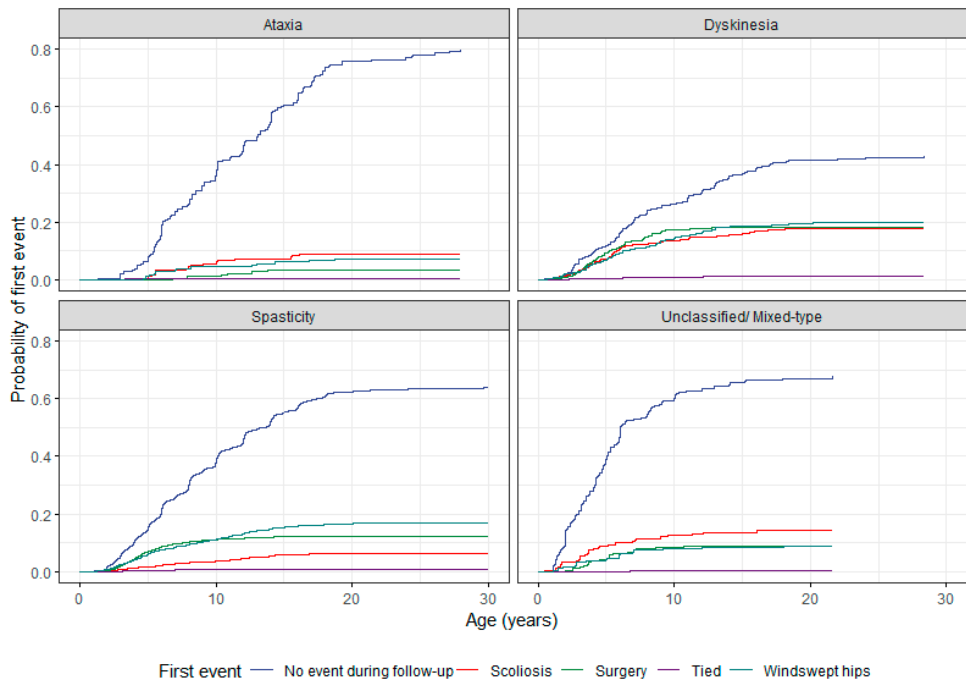


Figure 10. Probability of first event by CP subtype

Discussion

With improved neonatal care, maternal health and medical technologies the number of children with CP has reduced in Europe (17). However, CP remains the most common childhood onset disorder causing lifelong motor impairment (114) and continued use of healthcare services (115). Therefore, to facilitate participation in everyday activities and good quality of life it is vital that we understand typical presentation and associated risk factors. While most research of children with CP focuses on children at GMFCS levels I to III, this thesis includes children at all levels of motor function including those at GMFCS levels IV and V. This thesis focuses upon the sitting, lying and wheeled mobility of children with CP and how these are associated particularly with posture, postural ability, ROM, deformities, contractures, and pain.

The overall aims of this thesis were to increase knowledge of posture, postural ability, deformities, contractures, and pain in children with CP in supine and sitting (*Study II-III*); to determine risk factors for not independently using a wheelchair (*Study I*); and identify whether scoliosis or WSH occurs first (*Study IV*).

The sample of children was taken from the CPUP registry, providing access to children with CP in Sweden. With $\geq 95\%$ of all children with CP reported (110) into this large database it is expected that these results are representative of the total population. Results from the cross-sectional studies *I-III* offer understanding of prevalence and associations at a specific point of time. Therefore, none of these studies allow determinations of causality. In contrast the longitudinal study *IV* allows exploration over time in relation to the development of scoliosis and WSH. Further, studies *I-III* are the first to explore associations between the specific variables in a large cohort of children with CP.

Wheelchair use and independent wheeled mobility

Children with higher GMFCS levels have more severe mobility limitations and generally use a wheelchair for all their mobility needs (116). In Sweden children are provided with assistive devices such as a wheelchair, as a free loan, following assessment and prescription by a therapist. As the child grows or their needs change, the wheelchair is adjusted or replaced, so that it continues to meet their needs and enables mobility for the child.

We found that 1 in 4 children aged 0-11 years with CP used a wheelchair for indoor mobility and more than 1 in 3 for outdoor mobility. The majority who had a manual wheelchair were not able to independently self-propel. Having poor manual ability was found to be the greatest risk factor for not independently self-propelling, and more so for indoor than outdoor mobility. At the time this was surprising, as one would expect indoor manual mobility within smaller environments to have facilitated greater independence. However, work by Sonenblum and Sprigle (117) investigating the everyday use of manual wheelchairs the within-seat and pressure relief behaviours of adult manual wheelchair users with primarily spinal cord injury may shed some light in explaining this result. They found that these active, independent, manual wheelchair users completed many small distance bouts of activity within the home, and that these short bouts require much effort to overcome the initial inertia to move the wheelchair. Although a different client group, the continuous effort throughout the day might prove tiring for our children with CP, and hence contribute to fewer being independent manual wheelchair users.

It is also important to recognise that many children who need a wheelchair for their mobility often have postural challenges, especially maintaining sitting balance and postural control against gravity (118). Interestingly, Reilly et al. (119) have highlighted in their study with children with CP aged 10-14 years that maintaining postural control can be an attentionally demanding task. Further, whilst they simultaneously engage in another cognitively demanding task requiring them to dual-task, such as wheelchair use, their balance and postural control may be further compromised and lead to postural instability. Instability, lack of postural control, or even having postural asymmetries will mean that the child will require additional postural support (120) provided through the wheelchair seating in order to have stability in sitting. Lacoste et al. (121) found that a majority, 89% of manual wheelchair users, in a cohort of children with CP 8-18 years, became unstable in sitting when self-propelling, supporting our hypothesis that postural instability may be another reason for children not self-propelling in our study. This lack of stability may explain why so many of the children who used a manual wheelchair remained dependent and unable to self-propel. This level of detail on type of seating and support used within the wheelchair was not available to us, as not routinely reported into the national registry.

Addressing these postural needs and providing balance for the child can be complex and often requires expert understanding and provision of wheelchair seating (122). Indeed Ekiz et al. (123) in their study examined the appropriateness of wheelchair provision for children with CP in Turkey and found that over 80% of the children had inappropriate prescriptions. This raised concerns for sitting balance, upper limb functioning, postural alignment and risk of developing contractures and deformities. Further, the lack of postural control and stability in sitting can in turn make effective use of upper limbs challenging for the child. Without the added postural support in sitting, upper limb function may be restricted

(121, 124), and self-propelling difficult to achieve (121). The result being that children become more dependent on carers to push them, as we found in *Study I*.

In contrast with manual wheelchair users, we found that fewer children used a power wheelchair for their mobility, but when they did 75% drove independently. Further, children with GMFCS levels IV-V are known to require the use of mobility devices (20) so providing them with a power wheelchair could facilitate their independence with mobility (125, 126). It is somewhat surprising that the use of power wheelchairs remains under-used (127) and that more children were not provided with power wheelchairs, especially since we found that higher GMFCS level and poor manual ability were risk factors for not using a manual wheelchair independently. However, it is important to recognise that power wheelchairs are often large, heavy, and require the use of wheelchair adapted vehicles for transportation. Information on available transportation was beyond the scope of our study at this time, but would be important to consider in future work, and whether access to wheelchair accessible transportation is a barrier or facilitator to greater wheelchair use by the children. Additionally, Kenyon et al. (128) have suggested that some children are not afforded the opportunity to access power wheelchairs based upon the child's behavioural, cognitive, or physical factors. In our study we were unable to examine if cognition or vision were risk factors for not self-propelling or independently driving their wheelchair, as there was insufficient data on this variable.

An additional consideration for children not gaining wheeled mobility independence is that often parents can be reluctant to try or accept power wheelchairs (127), perceiving it as having 'given up', or accepting their child's level of disability or impairment (125). Yet, once access methods and the process of learning to drive are correctly individualised (126), and parents experience the freedom power mobility can offer the child (125) and the increased autonomy, they are much more amenable to its use.

Further, there remains a growing body of evidence highlighting how using power mobility can positively impact play skills, cognitive development, psychosocial skills, and participation (120, 129, 130) of children. In addition, Ragonesi et al. (131) have also suggested that power mobility may enable greater independence when compared to manual users. Our study supports this finding, and that the provision of power mobility should be given fuller consideration for young children with CP.

Posture and postural ability

The main finding of *Study II* was that postural asymmetries were present in over half of the 2,735 children in both sitting and lying positions (60.2% sitting and 53.6% in supine); and that they occurred across all age-groups, and GMFCS levels. Similarly, to Rodby-Bousquet et al. (50) in their study with young adults, and

Ágústsson et al. (113) with adults, we found an association between postural asymmetries and gross motor function. Children were more likely to have postural asymmetries as their gross motor function decreased. Specifically, children with GMFCS level V were more likely to have severe postural asymmetries in both supine and sitting positions. We did, however, find the presence of postural asymmetries for some children with lower GMFCS levels which was not expected as these children can move and change position. Therefore, it is important that children of all GMFCS levels are reviewed fully to ensure habitual patterns of postural asymmetry do not become established, regular changes in position facilitated, and the use of orthotic devices considered to prevent or reduce deformities and contractures (72, 132).

We found that in children with postural asymmetries there were differences in not only how many, but also, which body segments were involved as GMFCS levels increased. Children with lower GMFCS levels tended to have milder postural asymmetries involving fewer body segments and more often the feet or arms. Those children at higher GMFCS levels had involvement of more body segments, affecting the whole body. Furthermore, when considering sitting position, the postural asymmetries tended to involve the trunk and pelvis more often, whilst in supine it was the upper and lower extremities. Having postural asymmetries affecting the trunk and pelvis in sitting may contribute further to our understanding of why so many children using manual wheelchairs in *Study I* were dependent. This perhaps warrants further investigation as another possible risk factor for not being an independent wheelchair user.

Postural asymmetries in supine and sitting positions were found to increase as age increased, which has been reported by others (50, 133). However, we found postural asymmetries were present in the younger age-groups too, suggesting that for some children, these can start early in childhood. This supports work by Porter et al. (90) and Fulford and Brown (40) who have proposed that postural asymmetries may indeed derive early in infancy, and may be influenced by immobility. Porter et al. (90) report an association between the child being placed in an asymmetrical lying position in the first year of life, and the development of postural deformities. Further, they note that non-ambulant children are at even greater risk of having asymmetric postures and recommended preventing postural asymmetries from becoming an established pattern for the child. Indeed, we too found that the risk of having postural asymmetries increased as the ability to change position in supine and sitting decreased. In fact, the risk of having postural asymmetries in supine or sitting doubled if the child was unable to change position.

Having a habitual asymmetric posture for prolonged periods of time is believed to result in soft tissue adaptation, and the development of contractures and deformities (40, 50, 90, 134, 135)(Figure 11). The combination of having an asymmetric posture and not being able to change position may lead to the need for more postural supports in sitting and lying (134), compromised participation in everyday activities and reduced quality of life for the young person.

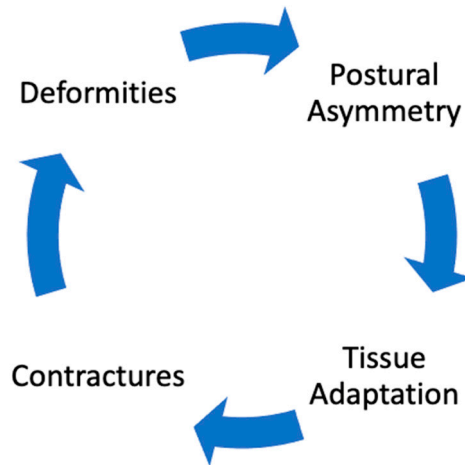


Figure 11. Cycle of postural asymmetries and development of deformities

For ease of management of the PPAS data we organised the scores and ratings into four categories for both postural asymmetries and for ability to maintain or move in/out of supine and sitting positions. To minimise assessment errors in completing the PPAS with the children, therapists require training in its completion (46), although it has been shown to have excellent inter-rater reliability even for untrained professionals (5).

We found a pain prevalence of 39% for the children, and a clear association between having postural asymmetries and having pain. Additionally, children had double the risk of having pain if they had severe postural asymmetries in supine or in sitting. We also discovered that children not able to change position in supine or sitting also had an increased the risk of having pain. Although there are variations in the prevalence and associations reported, there has been increased acknowledgement that children with CP may experience pain. Prevalence of pain is comparable to the findings of Alriksson-Schmidt et al. (111) 32.4% children aged 1-14 years, and Westbom et al. (136), and Jacobson et al. with 32-33% of young adults (137). Westbom et al. (136) who reported 37% children with CP as having reports of pain and identified locations of this pain. Eriksson et al. (138) reported a prevalence of 44% of children with pain, which affected their performance of daily activities or sleep. They also reported the risk factors for having pain as being female, older, or having higher GMFCS levels. In a multi-centre European study, the pain prevalence was reported as 74%-77%, much higher than we found. However, they only focused on 13- to 18-year-olds, and whether they had pain within the previous week. Additionally, they used the Bodily Pain and Discomfort items from the Child Health Questionnaire (139), enquiring if the location, circumstances, and if the child had pain during therapy, whilst we merely asked for the presence of current pain, and included both child and caregiver proxy reports of

having pain. Although there could be variations between child and caregiver identification of pain, when the child is very young, or has communication difficulties, caregiver reports have been acknowledged as an acceptable method of gathering this data (140).

Pain negatively influences participation in everyday activities (101, 141), leisure activities (142) and quality of life (140, 143), and can also create anxiety for children (144). Therefore, it is vital that we address pain (136), and one possible method may be to reduce the occurrence and development of postural asymmetries for children in CP.

Deformities and contractures

Deformities and contractures of the spine, hips and knees were present across all GMFCS levels and age-groups, including the very young. In our study knee flexion contractures were most frequent, followed by scoliosis; prevalence of all deformities and contractures was found to increase as age and GMFCS level increased. Although we did not examine hip displacement specifically, it is already a given that hip displacement in children increases with increased GMFCS levels and this is now used as a foundation in the development of hip surveillance guidelines in many countries (68, 145). In Clodt et al. (84) study with 3,045 children aged 0-15 years they reported a slightly higher prevalence of 22% knee contractures compared to our 19.2%, however, they included a cut off range of $\geq 5^\circ$ passive knee extension compared to our use of $\geq 10^\circ$. Similarly, they also identified that prevalence increased with age and GMFCS level. Persson-Bunke et al. (52) reported the same prevalence of moderate or severe scoliosis in children aged 4-18 years moderate or severe scoliosis as we found, and that this increased with age and GMFCS level. Willoughby et al. (68) reported a higher prevalence of 41% of scoliosis compared to our work, however, they examined scoliosis only in young people with skeletal maturity aged 16-29 years whilst we had all children 0-18 years who were still developing. They too reported associations with age and GMFCS levels. One explanation for their higher prevalence of scoliosis is that they used a lower cut off value of Cobb angle $\geq 10^\circ$ and had older children.

Further, in our study there were very few children (0.2%) who had a hip dislocation (MP of 100%) which was heartening; and lends support to the success of the surveillance programme in early monitoring and the prevention of hip dislocations for children with CP (106, 146, 147). In contrast, Aroojis et al. (105) report a prevalence of between 10-15% for hip dislocations for children with CP totalled across all countries with and without hip surveillance programmes. They also found that children regardless of sub-type presented with similar rates of hip dislocation, but rather they advocate for the use of a proactive approach to care using

early monitoring and hip surveillance to reduce this incidence, as is provided in Sweden for children with CP.

Another possible explanation for having lower prevalence of deformities and contractures in our population may be that all children aged 0-19 years avail of free of charge access to healthcare services in Sweden. This includes assessment, and treatment with multi professional paediatric habilitation teams, healthcare professionals, surgeries, prescription, and free loan of assistive technology devices (148) as assessed to be needed to meet their individual needs. In combination with the national surveillance programme these children are regularly monitored, receive early interventions, with the aim of reducing risk of deformities and contractures from occurring (100), or managing them early.

It is important to reiterate that generally children are born with a ‘typical’ spine, hips, and knees. Our finding that deformities and contractures were present across all age-groups raises concerns that some children may already have established these early in life, potentially impacting on all aspects of daily life. Porter et al. (90) also expressed a similar concern that postural deformities may occur early in life for children with severe motor impairment (GMFCS level V). They reported an association between the development of deformities and the child having an asymmetrical position in lying during its first year of life and explored the resultant direction of the deformities for the children.

Our work adds to the existing body of literature whereby Fulford and Brown (40) and then later Rodby-Bousquet et al. (50) with young adults, and Ágústsson et al. (41) with adults with CP, reported that lying for prolonged periods of time in an asymmetrical position could lead to the development of deformities and contractures. We found that children were more likely to have deformities and contractures if they were unable to change position.

In our studies we clearly found an association between having postural asymmetries, being unable to change position in supine and sitting, and with having deformities and contractures. In efforts to understand this relationship, brings us back to considering the potential negative influences of positioning as illustrated in Figure 11 (page 51), gravity and spending prolonged periods in one position as outlined by the Heuter-Volkman principle (Figure 7, page 27). We cannot change gravity, but as clinicians we can attempt to address the other elements of postural asymmetries whilst still reducible, through facilitating good postural alignment in supine and sitting, and frequent opportunities to change position throughout the 24-hour period. Early monitoring through surveillance programmes, enables identification of the children at risk of developing these deformities and contractures, and subsequently early and targeted interventions to be provided.

In our studies we used passive ROM to define the presence of WSH, adopting the methodology described by Persson-Bunke (113). This means that the WSH is objectively and consistently recorded and is not attributable to how the person was positioned in either supine or in sitting. Clinically, if someone has a pelvic obliquity, pelvic rotation, or limited hip or knee flexion these deformities and contractures can

directly impact on how the individual presents in sitting and may result in a 'windblown' or windswept appearance. Cloodt et al. (85) found that children with GMFCS levels III-V developed knee contractures first in the lower limbs. When in supine, knee contractures could tilt the legs to one side, and so present with a windswept appearance. This presentation may also vary depending on how the person is positioned or supported each time, and so give a less reliable measure of the incidence of WSH. Additionally, knowing how each author measures the presence of WSH is important in allowing comparisons in findings to be accurately made. For example, Ko et al. (149) use measurements of pelvis alignment about the transverse plane, taken from CT images to determine if a WSH exists. This method does not account for whether the WSH is a reducible postural presentation, or a fixed deformity; and is fully dependent on how the image was captured.

Further, we determined the child as having scoliosis when there was a moderate or severe curve on clinical examination using the Adams Forward Bend test, and/or on radiographic examination when there was a Cobb angle $\geq 20^\circ$. Again, the use of these cut-offs varies slightly from other researchers in that some have included mild curves, or Cobb angles $\geq 10^\circ$ (60). It is therefore likely our reported 10.5% will be a lower incidence of scoliosis. Indeed, this is evident when we look at several other studies investigating scoliosis. Yoo et al. (60) reported an incidence of 32.5% of scoliosis in their retrospective study with children and adults, McCarthy et al. (56) approximately 20% of children, Saito et al. (107) 68% in children with spastic CP, and Willoughby et al. (68) 41% in their study with young adults. As expected, each of these studies have a higher incidence of scoliosis as they had used a Cobb angle of $\geq 10^\circ$ to determine the presence of scoliosis; and some have studied slightly different populations, in terms of age, attending out-patient clinics, being institutionalised (53, 150), or having reached skeletal maturity (68).

Although Cobb angles of $\geq 10^\circ$ are categorised as scoliosis (56, 60, 63), children with flexible smaller curves and Cobb angles do not tend to receive treatment, particularly when they are still growing (63).

With deformities occurring above and below the pelvis at the spine and the hips, examination of whether scoliosis or WSH develops first will assist with knowing which deformity to address first, and for which children. Of the two deformities, we found that WSH occurred first; and that it occurred twice as frequent when compared to scoliosis. Our results with WSH occurring first were corroborated by Abel et al. (151) who reported 86%, and Letts et al. (78) who reported WSH occurred first in 86% and 68% of children respectively. They both reported higher incidences of WSH occurring first. This may be because they both had smaller samples; or only children who were about to have surgical intervention (151), or those with spastic CP (78). Further, it is also possible that they had higher incidences of WSH first as both studies were completed before the introduction of hip surveillance programmes targeted at reducing hip dislocations in children with CP.

We also examined CP sub-type in relation to whether scoliosis or WSH occurred first in a total population of children. Our study adds an important contribution to

the existing body of knowledge as much of the published literature has focused primarily on spastic CP. We found that children with dyskinesia and spasticity developed WSH first, with a higher incidence in the dyskinetic group. Children with dyskinetic CP also had the highest occurrence of scoliosis despite there being considerably more children with spastic CP in the whole sample. This finding differs from previous findings by Bertocelli et al. (67) who report spasticity to be a risk factor for having scoliosis. Their study had a much smaller sample size of 70 children, who were older at entry into their study, and were attending a specialist unit for spinal treatment, which may have introduced a selection bias within their sample. Furthermore, Pettersson et al. (66) identified risk factors for having scoliosis as being female, having epilepsy, GMFCS levels IV-V, and having limited knee flexion. Although our longitudinal study found that more children had WSH first when compared to scoliosis, we were able to agree that children with GMFCS level V, in addition to children with dyskinetic CP appeared to have a greater risk for having both scoliosis and WSH and did so at a lower age.

One possible explanation for children with dyskinetic CP presenting with the most scoliosis as a first deformity compared to the other sub-types may be related to these children often having difficulty maintaining position and presenting with trunk instability against gravity. Together this can create a vulnerability to adopting an asymmetric trunk presentation, leading to rotation, and ultimately scoliosis deformity as proposed by the Heuter-Volkman principle (Figure 7, page 27). Further, similar to Hägglund et al. (62) we found that children with higher GMFCS level were also more likely to develop scoliosis, and it is these children with lower gross motor function who typically have difficulty changing or maintaining a position. Moreover, like children with dyskinetic CP, children with GMFCS level V developed both WSH and scoliosis more often as a first deformity and did so at a younger age. This may again, relate to being in an asymmetric posture for a sustained period, and with not being able to move or change position. This finding further reinforces the need for these children to be carefully monitored, and as mentioned earlier, facilitated to have good postural alignment and frequent position changes in efforts to prevent these deformities from developing. Whilst children with dyskinetic CP should be provided with adequate supports in sitting and supine, such as bracing or adaptive wheelchair seating (152, 153), to enable them to maintain a stable aligned position.

Although there were more knee flexion contractures and scoliosis present for children in our *study III*, it was having WSH and hip flexion contractures which were associated with having pain. It is not clear why this was the case for the children, but it would appear important for reducing pain that hip deformities and contractures are prevented. Similarly in an earlier study with children aged 1-14 years, Alriksson-Schmidt et al. (111) reported a similar pain rate of 32.4% compared to our 38.3%, and later Eriksson et al. (138) reported 44% prevalence. Both these studies discovered pain to be more frequent in the lower extremities of feet, knee, and hips. Contrastingly, in their Danish registry study, Mehlenburg et al. (154) with

688 children, did not find any association between hip contractures and having pain. However, recording of the presence of pain in their study only focused upon presence in the lower limbs and not in general. This coupled with a smaller sample size and different child age range (5-12 years), may partly explain the different prevalence and lack of an association reported in their study. Reports of lower prevalence of hip associated pain in Swedish registry studies compared to the European SPARCLE studies (139, 155) could in part be attributed to the success of the surveillance programme in the prevention of deformities and contractures for the children (103); or could be influenced by CPUP being a national programme which is population-based with >95% children with CP reported into the database (110), and therefore reports on all children, and not just those children currently receiving healthcare provision (138).

Limitations

Study I

This study was cross-sectional in design so only possible associations influencing independent wheelchair mobility can be explored. As expected, hand function and upper extremity ROM were risk factors, however, only partially as many children with good manual skills still could not independently use their wheelchair. The grouping of variables and choice of cut-off values influenced the results. The reliability and validity of some of the hand functions is not fully established for children with CP.

The CPUP database lacks specific details on wheelchair model or configuration, or whether postural seating was used within the wheelchair. These factors can contribute to having core pelvic and trunk stability allowing greater upper extremity function and being able to efficiently reach the push-rims for self-propulsion or the controls on a power chair. Data was incomplete for cognitive and visual abilities and there was a lack of detail on access methods for power driving. Each of these factors may also have influenced the child's ability to use a wheelchair independently.

Study II

A limitation is our definition of pain prevalence, which included the presence of any current pain as reported by either the child or their proxy. This assumed that the primary caregiver can accurately report the presence of pain for the child. Further, this study reported the association of postural asymmetries and postural ability with pain, but not severity or location of the pain. As our study is a cross-sectional design no causality can be established between postural asymmetry, postural ability and the presence of pain.

Study III

A limitation was that to interpret the data for scoliosis, hip and knee flexion contractures cut-off values were used. This may have meant that our reported incidences were slightly lower. For scoliosis two categories of 'none' and 'moderate to severe' were used. These categories were selected as typically only children reported in the physiotherapy clinical examination as having moderate or severe scoliosis are referred for radiographical examination. Further, mild curves are only visible on forward bending examination, are not visible when in an upright position, and do not differ from no curve at radiographic examination. Persson-Bunke et al. (71) reported that only moderate curves visible in both forward bending and upright correspond to a scoliosis at radiographic examination with a Cobb angle of 20°.

To determine the presence of hip and knee flexion contractures a loss of passive hip and knee extension of $\geq 10^\circ$ ROM was used. Although this might result in some smaller range flexion contractures being excluded, it reduced the risk of measurement error by clinicians, and only included values that affect the biomechanical alignment of the joints and prevent straightening the legs.

Study IV

The GMFCS assessment was not available when the CPUP follow-up program and registry was started, so those children born 1990-1992 did not have a GMFCS classification at entry. However, the GMFCS level and CP subtype reported closest to the 5th birthday was recorded. Another limitation is that some children may not have had a long enough follow-up period to indicate if they developed either scoliosis or WSH.

As far as possible the date of assessment was used to determine which deformity developed first when they occurred within the same year. We included moderate and severe spinal curves on clinical examination, or those with $\geq 20^\circ$ Cobb angle on radiographs to indicate scoliosis. This was because moderate and severe curves at clinical examination show a high specificity and sensitivity with Cobb angles $\geq 20^\circ$ (71). This helps ensure that only children at risk of scoliosis progression are referred for radiographic examination.

Another limitation is that most children had the sub-type of spastic CP and much fewer in the other sub-types; and further the distribution of each sub-type differed across the GMFCS levels. Few children with dyskinetic CP were ambulators, with most having higher GMFCS levels, whilst for spastic CP the reverse was true with the majority being ambulators, and few having higher GMFCS levels. The impact of this distribution warrants further investigation.

As discussed on page 31 under registry-based studies, every effort has been made to minimise reporting and measurement errors. This has included annual registry data cleaning, local clinical training in assessment completion protocols, and data validation during our data analysis for each study.

Clinical implications

This thesis provides new knowledge that can enhance our clinical practice with children with CP.

There is a growing body of evidence that manual wheelchairs may not be the optimal solution for many children with CP to attain independent wheeled mobility, especially outdoors. Subsequently, there should be greater access to power wheelchairs for these children, and at earlier ages, to allow them maximal opportunities for independent wheeled mobility.

An association has been reported between having postural asymmetries in sitting and supine positions with having spine, hip and knee deformities and contractures. Therefore, postural assessments should be included in surveillance programmes, enabling early identification and treatment. Clinicians should address postural asymmetries whilst they are still correctable. Interventions should include the early introduction of postural management programmes of care as postural asymmetries have been detected across all age-groups of children, and GMFCS levels. Such interventions may include postural support and/or correction through positioning, orthotic devices (such as spinal braces, knee-ankle-foot-orthoses), wheelchair seating, customised seating, and night-time positioning to optimise function and minimise the risk of secondary complications.

Through the provision of accurate information of the relationship between prolonged asymmetric postures in supine lying and sitting with having deformities and contractures, we can support synergistic decision making by families in their postural management choices and the impact of these on posture, participation, and caregiving for their child.

Additionally, with finding an association between postural asymmetries, the inability to maintain or change position, deformities, contractures and having pain, clinicians should be made aware that for approximately one third of children there is an increased likelihood of having pain. Having an awareness of which deformity comes first will further assist with the targeting and prioritising of interventions for children with CP.



On a personal level, when I consider the most recent referral to our service, a three-year-old boy with CP I am very mindful of our research findings in planning his intervention alongside his family.

He is a young child requiring support in sitting to remain balanced and upright against gravity; and is unable to independently change his position. Consequently, he is at risk of lying or sitting in an asymmetric position for prolonged periods of time. Therefore, he is potentially at risk of developing contractures, deformities, having pain, and not being able to independently use a manual wheelchair should he need one in the future. Currently he is in a buggy which his parents use to push him around.

To prevent this child developing the same level of non-reducible deformities and contractures that Ruth now presents with as a 17-year-old with CP, I will strongly advocate for the careful management of his posture throughout the 24-hour period, regular monitoring of his hip ROM, spine, and pelvis alignment, as well as early access to assistive technology devices that can be adjusted to his postural needs, and orthotics. It is hoped that this will help protect his posture and enable him to be functional and independent within his community.

Conclusions

Overall aims of this thesis were to increase knowledge of posture, postural ability, deformities, contractures, and pain in children with CP in supine and sitting (*Study II-III*); to determine risk factors for not independently using a wheelchair (*Study I*); and identify whether scoliosis or WSH occurs first (*Study IV*).

Data for these studies came from the national registry and follow-up programme for cerebral palsy (CPUP) in Sweden. This registry has >95% of all children in Sweden with CP reported into it, and therefore provides access to a total population of these children. *Studies I-III* were cross-sectional in design so can facilitate reporting of associations between variables and not causation, whilst *Study IV* was longitudinal allowing us to monitor the children over a specified time period.

We found 1 out of 10 children could self-propel their manual wheelchair, whilst 3 out of 4 of those with a power wheelchair could drive independently. Poor hand function and gross motor function were the greatest risk factors for being unable to use a wheelchair independently. Over half of the children had postural asymmetries in supine and sitting positions, whilst 10.5% had scoliosis, 8.7% windswept hip deformity, 19.2% knee and 6.6% hip flexion contractures; and almost 4 in 10 children had pain.

Postural asymmetries increased with age and GMFCS-levels and doubled the likelihood for the child to have pain. Whereas having severe postural asymmetries increased the likelihood of having scoliosis (OR 9.1), WSH (OR 5.7-8.8), hip (OR 6.7) and knee flexion contractures (OR 12.2). The likelihood of having pain increased if the child had hip flexion contractures (OR 1.5) or WSH (OR 1.6). Those children unable to maintain or change position were more likely to have postural asymmetries in supine (OR 2.6-7.8) or in sitting (OR 1.5-4.2).

Further, there was a higher incidence of WSH (16.6%) as the first deformity than scoliosis (8.1%). WSH developed first in more children with lower gross motor function, and with dyskinetic (20.0%), or spastic CP (17.0%), whereas scoliosis developed in more children with GMFCS level V (19.8%) or dyskinetic CP (17.9%).

As clinicians it is vital that attention is focused on preventing or reducing postural asymmetries in supine and sitting for children with CP, where they spend most of their time; to reduce the risk of deformities and contractures of the spine, hip, and knee, and of having pain. It is also important that power wheelchairs are considered early as a possible means of facilitating independent mobility for these children. The totality of this cohort reporting on all age groups and GMFCS levels makes the results more generalisable to other populations of children with CP; and to those

with access to similar health care systems, whereby healthcare provision is free of charge for children.

In summary

Postural asymmetries in supine and sitting were present in over half the children, were associated with having WSH, scoliosis, hip and knee flexion contractures, and pain. Children unable to change position were likely to have postural asymmetries. WSH occurred first more often than scoliosis. Few children were able to self-propel a manual wheelchair independently, whilst most with a power chair were able to drive independently.

Future research

Questions that should be examined in the future include:

- 1) To investigate further whether children who acquire WSH or scoliosis first go on to develop other deformities or contractures.
- 2) To explore which assistive technology devices and dosages of use are being used, and are effective in managing postural asymmetries for children with CP.
- 3) To examine the use of posturally supportive seating in wheelchairs on the ability of children to independently self-propel a manual wheelchair.

Sammanfattning, summary in Swedish

Övergripande syfte med detta doktorandprojekt var att öka kunskapen om position, postural förmåga, felställningar, kontrakturer och smärta hos barn med CP i ryggliggande och sittande (*Studie II-III*); att identifiera riskfaktorer som hindrar självständigt förflyttning i rullstol (*Studie I*); och att undersöka om skolios eller windswept höftfelställning (WSH) inträffar först (*Studie IV*).

Data för dessa studier baseras på det nationella registret och uppföljningsprogrammet för cerebral pares (CPUP) i Sverige. Detta register följer över 95 % av alla barn i Sverige med CP och ger därmed tillgång till en totalpopulation av dessa barn. Studie I-III var tvärsnittsstudier för att analysera samband mellan olika variabler, medan studie IV var longitudinell och gjorde det möjligt för oss att följa barnen under en längre tidsperiod.

Vi fann att endast 1 av 10 barn kunde köra sin manuella rullstol själv, medan 3 av 4 med elrullstol kunde köra självständigt. Nedsatt handfunktion och grovmotorisk funktion var de största riskfaktorerna för att inte kunna köra sin rullstol självständigt. Över hälften av barnen hade asymmetrier i liggande och sittande, medan 10,5 % hade skolios, 8,7 % windswept höftdeformitet, 19,2 % knä- och 6,6 % höftflexionskontrakturer; och nästan 4 av 10 barn hade smärta.

Asymmetrier ökade med ålder och nedsatt grovmotorisk funktion klassificerad enligt Gross Motor Function Classification System (GMFCS) I-V. Asymmetrier fördubblade sannolikheten för smärta. Uttalade asymmetrier, som involverar hela kroppen, ökade också sannolikheten för skolios med nio gånger (OR 9,1), WSH mellan 5 och 9 gånger (OR 5,7-8,8), höftkontraktur över sex gånger (OR 6,7) och knäflexionskontrakturer 12 gånger (OR 12,2). Sannolikheten för smärta ökade också om barnet hade höftflexionskontrakturer (OR 1,5) eller WSH (OR 1,6). Barn som inte kunde bibehålla eller ändra position själva hade större risk för asymmetrier i ryggliggande (OR 2,6-7,8) eller sittande (OR 1,5-4,2).

Det var en högre incidens av WSH (16,6 %) än skolios (8,1 %) som första felställning. WSH utvecklades först hos fler barn med högre GMFCS-nivå och hos barn med dyskinetisk (20,0 %) eller spastisk CP (17,0 %), medan skolios utvecklades först hos en större andel barn i GMFCS-nivå V (19,8 %) eller med dyskinetisk CP (17,9 %).

Som kliniker är det viktigt att fokusera på att förebygga eller minska asymmetrier i liggande och sittande för barn med CP för att därigenom minska risken för felställningar, skolios, höft- och knäflexionskontrakturer och smärta. Det är också viktigt att vi redan tidigt överväger elrullstolar till barn med CP för att öka andelen barn som får självständig förflyttning. Data speglar en oselektad population av barn med CP i alla åldersgrupper och GMFCS-nivåer vilket gör resultaten lättare att generalisera till andra populationer av barn med CP som har tillgång till liknande hälso- och sjukvårdssystem, med fri sjukvården för barn.

Slutsatser

- Hälften av alla barn med CP har asymmetrier i liggande och sittande.
- Asymmetrier är associerade med ökad förekomst av windswept höftfelställning, skolios, höft- och knäflexionskontrakturer och smärta.
- Barn som inte kan ändra ställning själv har större risk för asymmetrier i liggande och sittande.
- Windswept höftfelställning utvecklas tidigare än skolios hos fler barn med CP.
- Få barn kan köra sin manuella rullstol självständigt, medan de flesta med elrullstol kan köra själv.

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