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WALKING IN ADULTS WITH SPASTIC CEREBRAL PALSY – THE RELATION TO PAIN, FATIGUE, GAIT AND BALANCE

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ABSTRACT

Background: Cerebral palsy (CP) is a disorder of movement and posture, and reduced gross motor function has been reported in several studies. Walking is an important function in everyday life, and important for independence. Studies on walking deterioration in adults with spastic CP are lacking.

Objectives: The main aim of this thesis was to investigate walking over a seven year period, to characterise and investigate the critical variables associated with walking deterioration, the changes in these variables over seven years, and to compare and characterise gait and balance in relation to walking deterioration.

Methods: Studies I and II were seven-year follow-up studies (N=149) using the same questionnaire as previously. Self-reported walking deterioration was assessed and compared. Pain, fatigue and gross motor function were compared over seven years, and the relationship between these variables and walking deterioration was examined (Study I). The relationship between number of pain sites and mental and physical components of Health-Related Quality of Life (HRQoL) was investigated, as well as pain site prevalence (Study II). For the inclusion to Studies III and IV, individuals in the critical age and sub-diagnosis for walking deterioration were identified. Three-dimensional gait analysis was used to obtain work of walking and kinematic gait variables in adults with spastic bilateral CP, <40 years in 2006 (N=16). The variables were compared between those reporting walking deterioration and those who did not. Gait was characterised across all (Study III). Number of falls was recorded. Balance confidence, fear of falling, balance ability were compared between those reporting walking deterioration and those who did not, and characterised across all (Study IV).

Results: Walking deterioration increased, and was most common in bilateral CP, especially in the 35-40 year age groups. Pain intensity and number of pain sites were unchanged, while pain frequency and HRQoL domain of bodily pain had worsened. Pain and physical fatigue scores were significantly higher in those reporting walking deterioration (Study I). Mental components of HRQoL did not correlate with the number of pain sites. Back, neck and foot/ankle were the most common pain sites (Study II). No differences in gait and balance variables were found between those reporting walking deterioration and those who did not (Studies III-IV). Across all, gait was characterized by increased anterior pelvic tilt, hip and knee flexion and deviating foot progression angle. Almost half was categorised as crouch gait (Study III). A high frequency of falls was found. Reduced balance in all subsystems was found. Postural responses and anticipatory postural adjustments seemed most affected (Study IV).

Discussion/Conclusion: Walking deterioration was associated with pain and fatigue. In contrast to the general population, psychological health was not associated with the number of pain sites, suggesting adequate coping strategies. The experience of walking deterioration was not explained by gait and balance variables, suggesting that other factors, possibly from all ICF domains, may be important. Balance was at the same level as elderly after fractures and the high number of falls may lead to activity and participation restrictions for these young individuals. These findings indicate a need for specialised and individualised follow-up programmes with a life-span perspective focusing on prevention of secondary consequences and of additional balance and gait deterioration.

Key words: Cerebral palsy, spastic, pain, fatigue, walking, gait, postural control, balance, kinematics, kinetics

SAMMANFATTNING

Bakgrund: Inom cerebral pares (CP), en störning av rörelse och posture rapporteras ofta nedsatt grovmotorisk funktion. Gången är en viktig funktion i det dagliga livet och vesentligt för oberoendet. Studier avseende hur gången försämras hos vuxna med CP fattas.

Syfte: Avhandlingens syfte var att undersöka gången över en sjuårsperiod, att karaktärisera och undersöka kritiska variabler relaterade till försämrad gång, och förändringar i dessa variabler över perioden, och att jämföra och karaktärisera gång och balans i relation till försämring i gångförmågan.

Metoder: Studier I and II är uppföljningsstudier (N=149), där samma enkät använts. Självrapporterad försämring av gångförmågan undersöktes och jämfördes. Förändring av smärta, fatigue and grovmotorisk funktion och sambandet mellan dessa variabler och gångens försämring undersöktes (studie I). Förhållandet mellan antal smärtställen, mentala and fysiska komponenter av hälsorelaterad livskvalitet (HRQoL), och prevalensen av smärta undersöktes (studie II). Till studierna III-IV blev individer i den kritiska ålder och sub-diagnos för försämrad gång identifierad (N=16). Gruppen med spastisk bilateral CP, <40 år i 2006 undersöktes med tredimensionell gånganalys för att erhålla kinetiska (arbetet) och kinematiska variabler. Dessa variabler jämfördes mellan de som rapporterade försämrad gång och de som inte gjorde, och karaktäriserades hos alla (studie III). Antal fall registrerades. Balanssäkerhet, fallrädsla och balanseförmåga undersöktes och jämfördes mellan de som rapporterade försämrad gång och de som inte gjorde, och karaktäriserades hos alla (studie IV).

Resultat: Förekomsten av försämrad gång ökade och var vanligast hos dem med bilateral CP, särskilt i åldern 35-40 år. Smärtintensitet och antal smärtställen var oförändrade, medan smärtfrekvens och HRQoL (domän kroppssmärta) försämrades över perioden. Alla smärt- och fatiguescorer var högre hos dem som rapporterade försämrad gång (studie I). Mentala komponenter av HRQoL korrelerade inte med antalet smärtställen. Vanligaste smärtställen var rygge, nacke och fot/fotled (studie II). Vare sig gånganalysens variabler eller de som rörde balansen visade några skillnader mellan grupperna (studier III-IV). Hela gruppens gång karaktäriserades av ökad bäckentippning framåt, ökad höft- och knäflexion och avvikande fotprogressionsvinkel. Nära hälften av alla gick med "crouch" gångmönster (studie III). Tretton ut av 16 hade mer enn fem fall sista år. Nedsatt balans framgick i alla balanstestets subsystem, särskilt inom posturala responser och anticipatoriska posturala justeringar (studie IV).

Discussion/Conclusion: En försämring av gången hade ett samband med smärta och fatigue. Psykologisk hälsa hade inget samband med antal smärtställen vilket skulle kunna bero på tillfredsställande coping strategier. Upplevelsen av att gången försämrats kunde ej förklaras av gång eller balansresultaten, vilket innebär att det är andra faktorer, möjligtvis från alla ICF domänen som är vesentliga. Balansen var reducerad tillsvarende äldre med höftfrakturer, och fallförekomsten var hög, möjligtvis ledande till reducerad delaktighet. Resultaten visar på behov av specialiserade och individualiserade uppföljningsprogram i ett livslångt perspektiv fokuserade på förhindring av sekundära komplikationer, liksom på att förhindra ytterligare försämring av gång och balans.

LIST OF PUBLICATIONS

This thesis is based on the following publications, which are referred to in the text by their Roman numerals:

- I. Walking function, pain, and fatigue in adults with cerebral palsy - a 7-year follow-up study. Opheim A, Jahnsen R, Olsson E, Stanghelle JK. *Dev Med Child Neurol* 2009; 51: 381–388.
- II. Physical and mental components of health-related quality of life and musculoskeletal pain sites over seven years in adults with spastic cerebral palsy. Opheim A, Jahnsen R, Olsson E, Stanghelle JK. *J Rehabil Med* 2011; 43: 382-387.
- III. Walking deterioration and gait analysis in adults with spastic bilateral cerebral palsy. Opheim A, McGinley J, Olsson E, Stanghelle JK, Jahnsen R [Submitted]
- IV. Balance in relation to walking deterioration in adults with spastic bilateral cerebral palsy. Opheim A, Jahnsen R, Olsson E, Stanghelle JK. [Physical Therapy, accepted for publication]

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ABBREVIATIONS

3DGA	3-dimensional gait analysis
ABC scale	Activities-specific Balance Confidence scale
BMI	Body Mass Index (body weight / body height ²)
BP	Bodily pain
CMC	Current Movement Capability
CNS	Central Nervous System
COM	Centre of Mass
CP	Cerebral palsy
FES-I	Falls Efficacy Scale – International
FMS	Functional Mobility Scale
FQ	Fatigue Questionnaire
FSS	Fatigue Severity Scale
GMFCS	Gross Motor Function Classification System
GP	General Population
GPS	Gait Profile Score
HRQoL	Health-Related Quality of Life
ICF	International Classification of Function, Impairment and Health
J	Joule
KI	Karolinska Institutet
MAMP	Maximum Achievable Movement Potential
MAP	Movement Analysis Profile
MCS	Mental Component Summary
MCT	Movement Continuum Theory
MDC	Minimal Detectable Change
N	Newton
Nm	Newton metres
OA	Osteoarthritis
PCS	Physical Component Summary
PMC	Preferred Movement Capability
ROM	Joint range of motion
SEM	Standard Error of Measurement
SF-36	Medical Outcome Study General Health Survey Short Form 36
SunHF	Sunnaas Rehabilitation Hospital
W	Work
W _{COM}	Potential energy of Centre of Mass Work

DEFINITION OF CONCEPTS

Walking deterioration	The self-reported experience of a worsened walking ability that has occurred over time.
Pain	An unpleasant sensory and emotional experience associated with actual or potential tissue damage, or described in terms of such damage (IASP 2011).
Fatigue	A sense of physical tiredness, and lack of energy, distinct from tiredness and sadness (Krupp et al. 1989).
Gait	Gait is the manner in which walking is performed (Malanga and DeLisa 1998). In this thesis, gait is often used interchangeably with walking, and also refers to how walking is performed in a controlled environment, such as a motion analysis laboratory.
Balance	The ability to control the body position in space for the dual purposes of stability and orientation (Shumway-Cook and Woollacott 2007). This is also called postural control, including both postural orientation and postural stability dimensions. The former includes the relationship between the body segments and between the body and the environment for a task. The stability dimension is the ability to control the centre of mass in relation to the base of support. In research into static balance, the latter dimension is normally studied. Balance, as understood and used in this thesis includes both dimensions, and postural control and balance are therefore used interchangeably.

1 INTRODUCTION

Cerebral palsy (CP) is the most common physical disability in childhood, affecting mainly motor performance, but also sensation, perception and cognition may be affected. The clinical appearance of CP is very heterogeneous, from the mildly to the severely impaired, with mental retardation and extensively reduced motor function. As the prevalence of CP is stable and more individuals with CP are now surviving well into adulthood, the number of adults with CP is increasing, so that they now outnumber children (Strauss et al. 2008). Some of the long-term consequences of living with CP are slowly becoming more evident, as research sheds more light on the health and mobility issues in adults with CP (Jahnsen 2004, Murphy 2010). However, the Cerebral Palsy International Research Foundation (CPIRF) still recognizes that the complications are “(...) largely under-recognised, undocumented and under-treated” (Cerebral Palsy International Research Foundation 2011). Further, a need to study methods to keep adults with CP ambulatory, as well as epidemiological studies on the magnitude of medical and orthopaedic complications, and health service research were identified as research priorities (Cerebral Palsy International Research Foundation 2011)(accessed 19/09/11).

Walking is an activity in itself, but also an important part of many other activities and participation in the community. The ability to sustain walking might be the crucial factor between being independent and being partly dependent on others in situations in everyday life. A functional reduction from being independent to becoming dependent on others for assistance has important consequences on the physical and psychological level for the individual, and has a considerable economic impact on society.

Reductions in walking and functional skills have been reported in other studies on adults with CP, but not all report walking deterioration. Some report stable function, some report deterioration, and some report cessation of walking. The factors that may describe the differences between those who report walking deterioration and those who do not, were the starting point for the present research projects. Factors associated with walking deterioration in previous studies were investigated in a 7 year follow-up study on walking function (Study I), pain (Studies I-II) and fatigue (Study I). Pain and its association with Health-Related Quality of Life (HRQoL) were additionally investigated in Study II. Based on these studies, the critical age and sub-diagnosis associated with walking deterioration were identified, and a group of people matching these criteria was investigated. Their gait was analyzed in order to compare differences in work of walking and overall kinematic gait deviation in Study III. As reduced balance was a frequently reported cause of walking deterioration, the relationship between balance and walking deterioration, as well as an overall characterisation of balance were investigated in study IV.

These studies highlight some of the critical factors that were associated with walking deterioration, and may therefore contribute to the body of knowledge concerning the development of walking in people with spastic CP in a life-span perspective. This may have implications for both the content and aims of rehabilitation programmes for this group.

2 BACKGROUND

2.1 CEREBRAL PALSY

2.1.1 Definition, causes and epidemiology

Cerebral palsy (CP) is a descriptive term, not a specific diagnosis, resulting from a non-progressive brain lesion occurring pre-, peri- or postnatally before the second year of life (Ingram 1984). A commonly used consensus-based definition of CP is: “an umbrella term covering a group of non-progressive, but often changing, motor impairment syndromes secondary to lesions or anomalies of the brain arising in the early stages of its development” (Mutch et al. 1992). A more recent definition also includes activity limitations and secondary health problems: “Cerebral palsy (CP) describes a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication, and behaviour, by epilepsy, and by secondary musculoskeletal problems” (Rosenbaum et al. 2007).

CP is caused by a disturbance of the development of the immature brain. There is a wide variety of developmental and acquired abnormalities that can cause the disturbance, and this can occur both in preterm and term infants, is most often cerebro-vascular, but can also be caused by trauma, infections and metabolic dysfunctions (du Plessis 2009). The incidence of CP in Norway is at about the same level as in the rest of Europe, which is 2.1/1000 live births/year, equivalent to 120-150 individuals/year (Andersen et al. 2008). With an average life expectancy of 70 years, the total CP population in Norway is estimated at approximately 8000 individuals. The number of adults above 18 years of age with spastic unilateral and bilateral CP, who are independent walkers without mental retardation, and thus those who are the main subjects of the present thesis, totals approximately 2000 individuals in Norway (Jahnsen et al. 2004b).

Surveillance of Cerebral Palsy in Europe (SCPE) classifies CP both according to both the most predominant feature (spastic, dyskinetic, ataxic), and the topographic representation (unilateral or bilateral) (Surveillance of Cerebral Palsy in Europe (SCPE) 2002). The subgroups of CP with mainly spastic impairments represents about 82% of the whole, divided into spastic unilateral, 33%, and spastic bilateral, 49% (Andersen et al. 2008).

The classification of CP according to SCPE is as follows:

Spastic CP is characterised by at least two of:

- Abnormal pattern of posture and/or movement
- Increased tone (not necessarily constantly)
- Pathological reflexes (hyper-reflexia or pyramidal signs e.g. Babinski response)
 - o It may be unilateral (hemiplegia) or bilateral (diplegia)

Ataxic CP is characterised by both of

- Abnormal pattern of posture and/or movement
- Loss of orderly muscular co-ordination, so that movements are performed with abnormal force, rhythm and accuracy

Dyskinetic CP is characterised by both of

- Abnormal pattern of posture and/or movement
- Involuntary, uncontrolled, recurring, occasionally stereotyped movements of affected body parts
- Dyskinetic CP may be either
 - o Dystonic CP, dominated by both hypokinesia and hypertonia.
 - o Chorea-athetotic CP, dominated by both hyperkinesia and hypotonia

The classification tree published by SCPE is presented in Figure 1.

Classification tree for sub-types of Cerebral Palsy

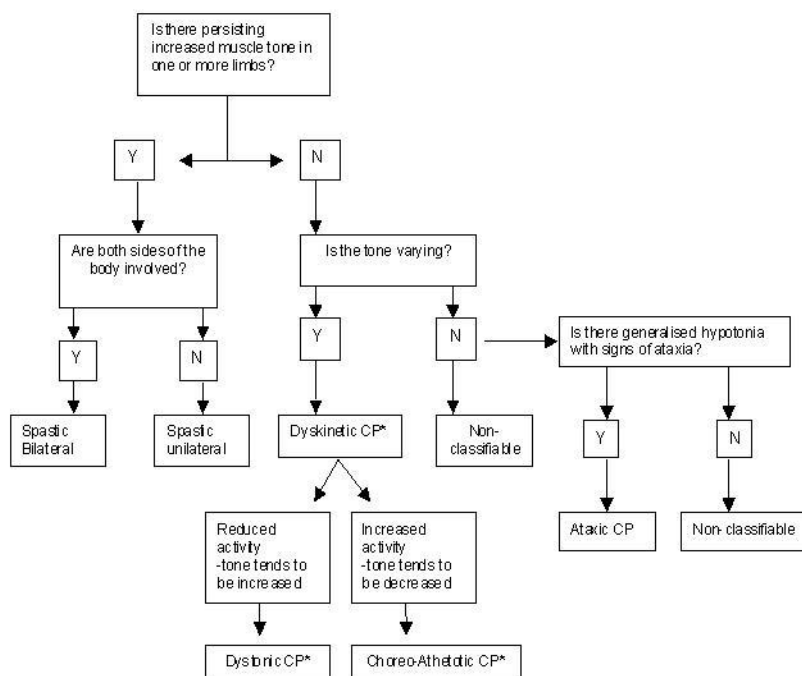


Figure 1. Classification tree for sub-types of Cerebral Palsy [Reprinted with permission from Surveillance of Cerebral Palsy in Europe, downloaded 10 January 2011 from: http://www-rheop.ujf-grenoble.fr/scpe2/site_scpe/decisiontree.php].

Spastic unilateral CP is often caused by a middle cerebral artery stroke (usually in term infants) affecting one brain hemisphere, while spastic bilateral CP is often caused by a brain injury referred to as peri-ventricular leukomalacia in preterm infants (du Plessis 2009). This latter lesion occurs most often in the white matter of the brain towards the posterior aspects of the ventricles, as well as adjacent to the frontal horns of the lateral ventricles, and involves both pyramidal and extra-pyramidal fibres. This injury leads to the typical clinical picture of spastic bilateral CP, where the lower extremities are more affected than the upper (Gage et al. 2009, du Plessis 2009). The pyramidal tracts are distributed primarily to the distal end of the

extremities. Therefore, selective motor control is more affected in the distal body parts than in the proximal. Bi-articular muscles are also often more affected than the monoarticular ones (Gage et al. 2009). For instance, in the posterior shank, the bi-articular gastrocnemius muscle is often more affected than the mono-articular soleus muscle (Gage et al. 2009), which can have important consequences for walking.

The brain lesion causes changes in the activity of the neural pathways, thus leading to the clinical picture of increased muscle tone or spasticity. This is caused by reduced inhibitory activity of the spinal motor neurons and vestibular nucleus. This causes increased stretch reflexes, muscle co-contraction, increased lower limb extensor contractions and reduced selective motor control and muscle weakness (Peacock 2009). The most commonly used definition of spasticity was proposed by Lance in 1980, as “a motor disorder characterised by a velocity-dependent increase in tonic stretch reflexes (muscle tone) with exaggerated tendon jerks, resulting from hyper-excitability of the stretch reflexes, as one component of the upper motor neuron syndrome” (Lance 1980) (p485). This definition has been challenged in recent years (Fleuren et al. 2010, Patrick and Ada 2006, Pandyan et al. 2005). However, as the clinical assessments of muscle tone, the modified Ashworth scale (Bohannon and Smith 1987), used in the present studies were based on this definition, it is used here, acknowledging that the resistance to rapid, passive movements includes both neural and mechanical components.

2.2 WALKING

Walking on two legs is one of the central motor functions that differentiate humans from the other primates. Walking is a highly regarded and valued motor skill, and our society is to a large degree designed on the assumption that people have this ability. The interest in and need for people with disabilities to be able to continue walking are therefore important to address. Normal walking is defined as: “a highly controlled, coordinated, repetitive series of limb movements whose function is to advance the body safely from place to place with a minimum expenditure of energy” (Gage et al. 1996). Gait in this thesis is understood as the manner in which walking is achieved (Malanga and DeLisa 1998). Fluent and flawless walking requires non-pathological functioning of the brain, nerve tract and motor units, as well as the muscles that work as levers of the normal skeletal system. This translates muscle actions into motions, and allows for propulsion. A normal central nervous system controls balance and coordinates step length and cadence in relation to the environmental challenges. Proprioceptive information from the skin and the joints are necessary for fine tuning and walking on uneven surfaces. Respiration and cardiovascular functions with sufficient reserves are necessary for walking endurance and increases with walking velocity. Injuries or dysfunctions in any of these systems can lead to walking difficulties (Gage et al. 1996).

The Movement Continuum Theory of Physical Therapy (MCT) (Cott et al. 1995) emphasises movements as essential, and as existing from the cellular level to that of the individual in society. Walking is an activity that may be affected by malfunctioning at any of these levels (Gage et al. 1996, Cott et al. 1995). At each level, MCT emphasises a maximum achievable movement potential (MAMP), which is dependent on the individual combination of biological, psychological and social factors. Reduced muscle strength and motor control or increased muscle tone may affect the MAMP, and lower the physiological reserves. MCT describes a preferred

and a current movement capability (PMC and CMC), which in normal circumstances are the same, but they are not invariable. For instance, the PMC may change due to individual choices, and the CMC may change for instance due to pain, fatigue, or aging (Cott et al. 1995).

2.2.1 Walking in children with CP

Independent walking in people with CP is generally achieved at a later stage than in typically developing children. In a recent Norwegian study (Andersen et al. 2008) children with CP were assessed for both fine (e.g. hand function) and gross motor (e.g. walking) skills (N=294). The median age of assessment was 6.9 years (range: 1.9-10.2). Across all subtypes of CP, there were 55% who had developed independent walking, while 16% walked with assistive devices, and 29% were unable to walk. Median age when the independent walkers started to walk was 22 months (range: 10-77), while those dependent on assistive devices started to walk at 36 months of age (range: 18-80). Among the children with spastic unilateral CP 95% walked independently, while in spastic bilateral CP 37% walked independently, 25% walked with assistive devices and 38% were unable to walk (Andersen et al. 2008). The development of gross motor functions has been found to stabilise and plateau at different ages (Rosenbaum et al. 2002), and those who were at Gross Motor Classification System (GMFCS) level I stabilised their gross motor development at a later stage, around the age of 5 years, while those at GMFCS level V stabilised earlier, at around 3 years of age (Rosenbaum et al. 2002). A recent population-based study of 562 children with CP in the south of Sweden, of whom 66% were spastic, found that walking performance increased up to seven years of age, and that the proportion of children who were able to walk independently on uneven surfaces was higher in each age group up to 18 years of age (Rodby-Bousquet and Hägglund 2011). As around 70% of all children with spastic CP can walk (Andersen et al. 2008), albeit in some cases with considerable difficulty, optimising walking function in a life-span perspective is important.

GMFCS is the most common system to classify gross motor function in children and youth in CP (Palisano et al. 1997). GMFCS is divided into five levels:

- Level I: walks without restrictions; limitations in more advanced gross motor skills,
- Level II: walks without assistive devices; limitations walking outdoors and in the community,
- Level III: walks with assistive mobility devices; limitations walking outdoors and in the community,
- Level IV: self-mobility with limitations; children are transported or use power mobility outdoors and in the community,
- Level V: self-mobility is severely limited even with the use of assistive technology.

The causal factors behind the gait problems in CP have been divided into primary, secondary and tertiary factors (Gage et al. 2009). Gage (Gage et al. 2009)(p107-128) states that pathological gait is a mixture of primary, secondary and tertiary abnormalities:

- Primary abnormalities (impaired balance, abnormal muscle tone and impaired selective motor control)

- Secondary abnormalities (impaired muscle and bone growth, and other tissue contractures leading to lever-arm dysfunction)
- Tertiary abnormalities (coping responses in the gait pattern as a consequence of the primary and secondary abnormalities)

The assumption behind this division is that the secondary and tertiary abnormalities are consequences of the primary and secondary abnormalities, respectively. This assumption also implies that treatment and therapy should primarily be directed towards the primary and then to the secondary abnormalities, and as a result, the tertiary abnormalities would correct themselves, as they are no longer necessary (Gage et al. 2009).

As the children with CP grow older, taller and heavier, the primary abnormalities of reduced motor control and increased muscle tone often lead to secondary musculoskeletal consequences such as joint contractures and reduced joint range of motion (ROM), muscle weakness and bony deformities such as tibial torsion, femoral anteversion, pes equinovarus or equinovagis and mid-foot break (Gage et al. 2009). These musculoskeletal abnormalities have been found to contribute to impaired walking and overall gross motor function, as well as to abnormal joint loading, and progress across childhood (Gage et al. 2009). Therefore, it is often necessary to correct these musculoskeletal abnormalities surgically after the gross motor function development has plateaued, and before skeletal maturation (Bache et al. 2003). The development of these deformities and their impact on walking into adulthood is largely unknown.

The main contributions to the increasing body of knowledge about gait in children with CP have come from developments in 3-dimensional gait analysis (3DGA). 3DGA has become an important tool in the interpretation and understanding of gait problems (Gage et al. 1996), and has become an important assessment tool both in preoperative decision-making, and in evaluating pre- and postoperative gait in children with CP (Sutherland and Davids 1993, Kay et al. 2000, Cook et al. 2003, Lofterod et al. 2007, Gage et al. 2009).

Early treatment of spasticity, together with a structured follow-up programme with the aim of preventing hip dislocation and severe contractures and deformities [Cerebral Pares UppföljningsProgram (CPUP)], has been implemented in Sweden since 1994, starting in the south of Sweden (Skåne and Blekinge), and now aiming to include all children with CP in Sweden. Hägglund et al. (2005) studied the effect of early spasticity treatment using either Intrathecal Baclofen, Selective Dorsal Rhizotomy (SDR) or Botulinum Toxin injections, together with the CPUP programme. The results showed a significant reduction in the number of orthopaedic operations to correct deformities or salvage dislocated hips, suggesting that early interventions to reduce spasticity might prevent the development of severe contractures and deformities, thus reducing the need for orthopaedic operations (Hägglund et al. 2005). The development of lower limb joint range of motion (ROM) from 2-14 years of age in children with CP in southern Sweden was reported by Nordmark et al. (2009), who found decreased ROM from 2 to 14 years of age. The reduction was generally more pronounced in those with more reductions in gross motor function. As the children grew older knee extension was reduced in those with spastic bilateral CP, and ankle dorsiflexion and hip external rotation were reduced in those with spastic unilateral CP (Nordmark et al. 2009). Decreased maximal ankle

dorsiflexion was found to be significantly related to spasticity in the gastrocnemius and soleus muscles, with higher degree of spasticity leading to greater reductions in maximal ankle dorsiflexion (Hägglund and Wagner 2011) .

The natural history of walking in children with CP during their growth has been studied by Johnson et al. (1997). They followed 18 children with spastic bilateral CP (4-14 years of age), over a mean period of 32 months, using gait analysis. Negative changes in walking velocity and stride length, as well as kinematic variables with reduced pelvis, knee and ankle joint excursions during walking were reported. This indicates a gradually worsened walking ability (Johnson et al. 1997). Negative changes in kinematic and spatio-temporal gait variables were also reported by Bell et al. (2002), who studied a group of 24 children with spastic unilateral and bilateral CP over a mean period of 4.5 years (from a mean age of 7.8 to 12.2 years). They also found reduction in hip, knee and ankle excursion, as well as decreased single support, reduced walking velocity and stride length (Bell et al. 2002). These changes in gait variables during growth were hypothesised to be caused by a discrepancy between skeletal and muscle growth. As muscle growth is not able to keep up with bone growth, this causes muscle shortness, contractures, bony torsions and reduced range of motion (Johnson et al. 1997). Bell et al. (2002) have included several other factors in addition to contractures that might be important for the observed deterioration, such as body weight, body weight/muscle strength ratio, spasticity, and compliance with physical therapy and bracing. A retrospective study by Day et al. (2007) showed that from 10-25 years of age the probability of stable or improved function were higher for those with better walking function, and the probability of losing walking ability was greater for those using a wheelchair.

2.2.2 Walking in adults with CP

When the individuals with CP become adults, new challenges arise related to education, work and family life. Compared to the number of articles about gait in children with CP, research on gait in adults is scarce, and the current knowledge of adult gait is limited. There are several reports on functional mobility deterioration in adults with CP (Balandin and Morgan 1997, Bottos et al. 2001, Andersson and Mattsson 2001, Murphy et al. 1995, Ando and Ueda 2000, Jahnsen et al. 2004b). In the Norwegian survey of the physical and social life situation of 406 adults with CP based on postal questionnaires, Jahnsen et al. (2004b) found deteriorated walking before the age of 35 among 22% of the people with spastic unilateral and in 52% of the people with spastic bilateral CP, most frequently between the ages of 15 and 34. Older age and severe neurological impairment were statistically associated with loss of walking ability. Reduced balance was commonly reported. The self-reported causes of deterioration of walking skills were pain, fatigue and reduced participation in physical activities (Jahnsen et al. 2004b).

In both spastic unilateral and bilateral CP some people experience a loss of walking ability. The extent of this loss is lower in spastic unilateral CP, probably because they have one body half that is not directly affected, but are compensating for the impairments in the other side. Jahnsen et al. (2004b) found that in spastic unilateral CP 2% had stopped walking in adult age, while in bilateral CP 12% had stopped walking, and 14 of 16 had stopped before the age of 45 years, using other means of ambulation, such as manual and electric wheelchair. Day et al. (2007) retrospectively studied a large group (N=5771) of adults with CP in California, from the age of 25 till

40. They found that the probability of losing walking ability was greater for those who walked unsteadily or with support and used a wheelchair at the age of 25 than for those also walking unsteadily, but not using a wheelchair, and almost none who initially walked well had lost walking ability (Day et al. 2007).

Longitudinal studies on the natural history of gross motor function and walking, and the associated factors such as pain and fatigue are few in number. Hanna et al. (Hanna et al. 2009) followed 657 children up to 21 years of age. They found that those at GMFCS levels III-V showed decreased gross motor function after the age of 7-8 years, while those at GMFCS levels I-II remained stable (Figure 1).

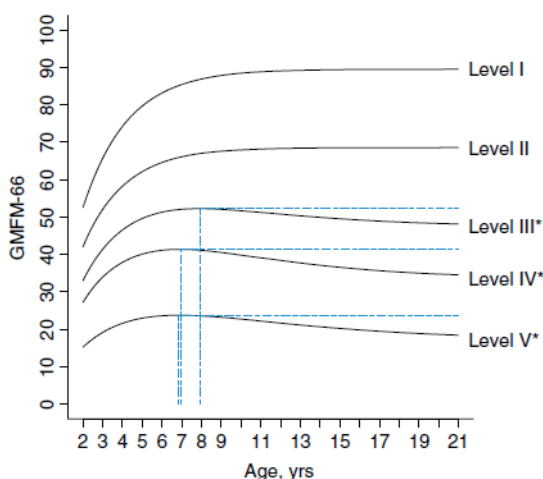


Figure 2. Predicted Gross Motor Function Measure (GMFM-66) motor scores as a function of age by Gross Motor Function Classification (GMFCS) level. *GMFCS levels with significant average peak and decline. Dashed lines illustrate age and score at peak GMFM-66. [Copyright 2009 Wiley. Used with permission from Hanna SE et al. Stability and decline in gross motor function among children and youth with cerebral palsy aged 2 to 21 years. *Dev Med Child Neurol*. 2009;51:295-302. MacKeith Press]

McCormick et al. (McCormick et al. 2007) studied the stability of gross motor function from childhood to young adulthood in a clinic-based study. They showed relatively stable GMFCS levels from childhood and up to a mean age of 22 years (weighted kappa: 0.895). Most of the changes were found to occur between levels I and II, and between levels IV and V, but no systematic changes were found. The positive predictive value was found to be 88%, meaning that for a person at GMFCS levels I and II at the age of 12 years, there is a 88% chance that s/he is at the same level, and has not deteriorated to levels III-V at the age of 22 years (McCormick et al. 2007). Sandström et al. (2004) found that in 16 out of 48 individuals the GMFCS level had deteriorated from adolescence up to a mean age of 33 years. The changes in GMFCS levels in adults is still an area which needs to be researched and this was therefore included as one of the aims of Study I.

The studies reporting functional deterioration in adults with CP are all cross-sectional studies (Balandin and Morgan 1997, Bottos et al. 2001, Andersson and Mattsson 2001, Murphy et al. 1995, Ando and Ueda 2000, Jahnsen et al. 2004b), except Ando

and Ueda (2000) and Day et al.(2007). Ando and Ueda reported that eight out of 90 people in their mid-thirties had a functional deterioration in walking over a five-year period (Ando and Ueda 2000). However, this study included only individuals who worked at specific community workshops, and the representativeness of this group compared to the total group of adults with CP can therefore be questioned. Day et al. (2007) showed that among those who initially walked well, but used handrails for stairs at the age of 25, about 30% had a decline in walking ability by the age of 40.

A few follow-up studies on adults with CP after surgical treatments have been published. Langerak et al.(2008) studied 13 individuals 20 years after SDR, and Gannotti et al. (2010) followed up a group of 23 individuals over 13 years (range 11-15) after single-event-multi-level surgery (SEMLS). None of these studies found deterioration in kinematic variables from adolescence to young adulthood. However, the latter studies have investigated smaller samples and individuals at a younger age (mid-late twenties) than in the studies by Jahnsen et al. (2004b), Andersson and Mattsson (2001), Ando and Ueda (2000) and Bottos et al. (2001). Individuals who have undergone SDR and SEMLS are also a selected group of individuals, making the comparisons with population and rehabilitation based studies difficult.

Most studies on walking function in adults with CP are based on self-reporting and not on instrumental gait analysis (3DGA). Detailed characterisations of kinematic and kinetic gait variables, as well as the categorisation of gait patterns in adults with CP are therefore almost totally absent. Only one study used 3DGA when describing gait in adults with CP (Abel et al. 1997). Twenty-two adults with CP, most of them with spastic bilateral CP (n=16) continued to experience active and passive joint motion abnormalities and diminished motor function. Gait abnormalities, lower extremity pain and radiographic abnormalities were prevalent findings that were related to the level of motor impairment (Abel et al. 1997).

The above findings on walking and its development over time in spastic CP show a gap in the current knowledge about adults' gait, which must be bridged, in order to improve therapy and development of lifespan models of rehabilitation and care for people with spastic CP. As a beginning, the changes in GMFCS levels and their relationship to walking deterioration were investigated in Study I. As the kinematic gait deviations, the work of walking, or balance ability in relation to walking deterioration is unknown, these relationships were investigated in Studies III and IV. Such information may be important in order to be able to identify those at risk of walking deterioration, and perhaps a later loss of walking. Relevant rehabilitation actions and physiotherapy interventions might have a better starting point, and may contribute to delay, or assist in a better coping with a deteriorated walking function.

2.3 PAIN

Pain has been defined by the International Association for the Study of Pain (IASP) as: "An unpleasant sensory and emotional experience associated with actual or potential tissue damage, or described in terms of such damage" (IASP 2011) (accessed 22/08/11). This definition clearly acknowledges pain as a subjective experience, links it to emotions, and makes it clear that pain can exist without tissue damage.

Different types of pain are often described based on the origin of sensory experience. According to IASP, neuropathic pain is, “pain caused by a lesion or disease of the somato-sensory nervous system.” Neuropathic pain can be divided into central or peripheral neuropathic pain, depending on the origin of the lesion or disease being in the central or the peripheral nervous system (IASP 2011) (latest access 22/08/11). Further, IASP describes nociceptive pain as, “pain that arises from actual or threatened damage to non-neural tissues, and is due to the activation of nociceptors. This term is designed to contrast with neuropathic pain. The term is used to describe pain occurring with a normally functioning somatosensory nervous system to contrast with the abnormal function seen in neuropathic pain” (IASP 2011) (latest access 22/08/11). Musculoskeletal pain, as studied in this thesis, may be classified as nociceptive pain, because the pain originates from non-neural tissue. In the definition of CP, it is stated that the motor disorders of CP are: “(...) often accompanied by disturbances of sensation, perception, cognition, communication, and behaviour, by epilepsy, and by secondary musculoskeletal problems” (Rosenbaum et al. 2007). Because of the disturbances of the sensory system, people with CP may also have neuropathic pain conditions. In the present studies no discrimination was made between neuropathic and nociceptive pain.

It is well known that pain sensitisation can take place in long-standing pain. Sensitisation has been defined by ISAP as: “increased responsiveness of nociceptive neurons to their normal input, and/or recruitment of a response to normally subthreshold inputs”, and can occur both in the central or in the peripheral nervous system, depending on the location of the neurons with increased responsiveness (IASP 2011) (latest access 22/08/11). Pain sensitisation is often associated with hyperalgesia, i.e. increased pain response on painful stimuli, and has been found in individuals with chronic pain (Giesecke et al. 2004). Neuropathic symptoms were found to be prominent features of chronic musculoskeletal pain, suggesting that these symptoms may represent an underlying sensitisation (Giske et al. 2009). The process of pain sensitisation can also take place in people with CP who experience chronic pain. Maslon et al. (2011) have studied a group of non-ambulant children with CP with dislocated hips, and found an increased density of nociceptor fibres in most of those with painful hips, and a correlation between the density of nociceptor fibres and subjective pain intensity. Thus, this study has demonstrated the presence of pain sensitisation (Maslon et al. 2011). Whether pain sensitisation also has taken place in ambulatory people with CP, who might have less pain intensity than the non-ambulatory with hip dislocations, has not been studied.

In order to understand the extent and impact of pain in adults with CP, it may be useful to briefly review some of the research on pain in adults from the general population (GP) in Norway. Musculoskeletal pain is common in the GP, and Natvig et al. (1995) showed the prevalence of chronic musculoskeletal pain (daily pain > 1 year) to be 15%, and that 15% reported no pain. The prevalence of chronic pain increased with age (Blyth et al. 2001). Gender differences in pain have been found both in the Netherlands (Wijnhoven et al. 2006) and in Norway (Rustoen et al. 2004). Pain at one site was found to be a predictor for the development of pain at other sites, and pain at additional sites increased the likelihood that the pain syndrome would become chronic (Croft et al. 2006). When pain is seen as a continuum from localised to widespread, the number of pain sites can be used as a simple measure of “widespread” pain (Kamaleri et al. 2008a, Croft 2009). In the GP, the number of pain sites was found to be associated with reduced overall health, sleep quality, female gender and psychological health (Kamaleri et al. 2008a), and was a strong predictor

of work disability (Kamaleri et al. 2008b). The relationship between the number of pain sites and physical and mental components of Health-Related Quality of Life (HRQoL) in adults with CP has not been studied previously, and was addressed in Study II.

2.3.1 Musculoskeletal pain in adults with CP

Musculoskeletal pain, in this thesis called pain, if not explicitly stated otherwise, was found to be common in adults with CP, with a prevalence of chronic pain (daily pain >1 year) of 28%, almost double that in the GP in Norway (Jahnsen et al. 2004a, Natvig et al. 1995). The findings from Jahnsen et al. (2004a) are consistent with other studies on pain in people with CP (Schwartz et al. 1999, Engel et al. 1999, Jensen et al. 2004). Malone et al. (2009) also found a significant negative correlation between the number of pain sites and reduced participation in activities resulting from pain. The prevalence of chronic pain in adults with CP varies from 18-84% (Andersson and Mattsson 2001, Turk et al. 1997). These wide variations are partly caused by differences in the selection of people with CP included in the studies, and the use of different definitions of chronic pain. Comparison between studies is therefore challenging. There is reason to conclude that musculoskeletal pain is more prevalent in people with CP than in the GP (Jahnsen et al. 2004a).

Jahnsen et al. (2004a) found that chronic pain in adults with CP was significantly associated with reduced physical role function, low life satisfaction and deterioration of functional skills. Pain may therefore have negative consequences on the degree of activity and participation in everyday life, and have a close association with walking deterioration, but the causal pathways are not clear. Pain may lead to an experience of deteriorated walking, such as shorter and more restricted walks. On the other hand, a deviating walking with increased muscle work, structural changes such as mid-foot break or rotational deformities in the long bones, and abnormal joint loading may also lead to increased pain. Both Andersson and Mattsson (2001) and Jahnsen et al. (2004a) found that pain was associated with reduced gross motor function and walking, and Murphy et al. (1995) found that joint pain was the main reason for giving up walking for people in their mid-forties. These findings indicate that pain may be an important factor for walking deterioration in adults with CP. This relationship was therefore investigated in Study I.

Back, neck and the weight bearing joints were found to be the most prevalent pain sites in several studies (Schwartz et al. 1999, Andersson and Mattsson 2001, Jahnsen et al. 2004a, Murphy et al. 1995, Engel et al. 2003). Whether this changes over time or is a consistent and stable pain characteristic has not been investigated. This was therefore investigated in Study II.

In a recent review article, Vogtle (2009) found limited evidence of interference of pain on activities and participation in CP, and the included studies showed variations in the number of individuals reporting pain interference. This might be due to the limitations in activities and participation already caused by the motor impairments in CP. Vogtle (2009) concludes that pain is a major problem that is consistently reported across several studies. The pain interventions seemed underutilised, and there was a need for both descriptive and experimental studies, as well as studies on pain incidence, sources and management (Vogtle 2009).

The pain in adults with CP may originate from a variety of body structures, and combinations of these. Muscle weakness, overuse, spasticity, stiffness and exertion can cause pain. The muscles in people with CP are often weaker than in typically developing children (Damiano et al. 2001). This may occur both because these individuals are often less active than their peers (Michelsen et al. 2009), and because of reduced agonist activation (Stackhouse et al. 2005). Increased muscle tone may cause pain and discomfort and lead to movement difficulties, both when the muscles act as agonists and antagonists. This leads to increased work of walking, as found in several studies in children with CP (Rose et al. 1990, Corry et al. 1996, Duffy et al. 1996, Johnston et al. 2004) and adults with spasticity (Mattsson et al. 1990).

Because of delayed gross motor development, abnormal muscle and loading forces and contractures, bony deformities are common in spastic CP. Three common bony abnormalities in spastic CP are increased femoral anteversion, tibial torsion (Gage et al. 2009), and hip dysplasia, which has been found to be more common in those with higher GMFCS levels (Häggglund et al. 2007). This may lead to abnormal joint loading, the ligaments and joint capsule might both be stretched in some directions and shortened and contracted in others. The joint cartilage may be strained in positions where it is not designed to take pressure, thus leading to early cartilage damage, and to the development of osteoarthritis (OA) (Carter and Tse 2009, Murphy 2009). Increased pain and OA changes may potentially take place in all joints with abnormal loading, and early development of OA has been found in CP (Carter and Tse 2009). The overall prevalence of OA in different age groups of people with spastic CP has not been investigated, but Boldingh et al. (2005) reported OA in 59% of a group with severe CP, and Cathels and Reddihough (1993) found that 27% in a group of 15-25-year-old people with CP had OA changes.

2.4 FATIGUE

The concept of fatigue may be understood in different dimensions (Widerstrom-Noga and Finlayson 2010). When fatigue can be attributed to the disease, such as the additional demands on the surviving motor units in polio, it has been called primary fatigue. Secondary fatigue encompasses many other factors, not directly related to the disease, such as cardiorespiratory functions, sleep problems, medication, nutrition, depression etc. The different dimensions of fatigue also include the duration (acute or chronic), experienced versus physiological, physical versus cognitive, as well as peripheral (muscles or peripheral nerves) versus central fatigue (CNS) (Widerstrom-Noga and Finlayson 2010). In the physiological sense, fatigue is the inability to develop and/or sustain muscle power output. Fatigue may also be related to the subjective experience of feeling tired, weak and in lacking in energy (Lerdal et al. 2005). Krupp et al. (1989) defined fatigue as: “a sense of physical tiredness, and lack of energy, distinct from tiredness and sadness”, and the Fatigue Severity Scale (FSS) is based on this definition. In this thesis, fatigue in adults with CP is understood according to the definition by Krupp (1989).

Fatigue was found to be common among individuals who are aging with chronic disabilities such as multiple sclerosis, spinal cord injury, polio, and in early acquired developmental disabilities such as CP (Chaudhuri and Behan 2004, Widerstrom-Noga and Finlayson 2010, Strax et al. 2010). Fatigue is an unspecific symptom which is frequently reported in the GP, and a prevalence rate of 11% of chronic fatigue (daily fatigue, >6 months) in the Norwegian GP has been found (Loge et al. 1998a). Fatigue

has been found to increase with age, women reporting higher levels than men, and those reporting diseases or current health problems report higher levels of fatigue than those without (Loge et al. 1998a). Higher prevalence rates of chronic fatigue in the GP have been reported in other studies, but these differences seem mainly to be attributed to the study samples (Chen 1986, Loge et al. 1998a, Lerdal et al. 2005).

2.4.1 Fatigue in adults with CP

In people with CP, fatigue has been found to have a higher prevalence than in the GP in Norway (Jahnsen et al. 2003b, Loge et al. 1998a). Fatigue in people with CP can be argued to be a primary fatigue due to reduced voluntary motor activation, but can also be argued to be secondary fatigue due to inefficient movement patterns, increased muscle tone, antagonist co-contraction, muscle weakness and reduced cardiorespiratory fitness. Fatigue was mainly related to physical functions like feeling weak and in lack of energy, and to a lesser degree related to mental functions like concentration, memory and to finding the correct word (Jahnsen et al. 2003b). In people with CP, fatigue was found to affect older individuals more than younger ones and females more than males. Further, fatigue was more prevalent in individuals with a self-reported moderate grade of CP (Jahnsen et al. 2003b). In a multiple regression analysis, fatigue was significantly associated with limitations in physical and emotional role function, deterioration of functional skills, bodily pain and low life satisfaction (Jahnsen et al. 2003b). The increased energy expenditure during walking in spastic CP (Rose et al. 1990), in addition to the reduced physical fitness (Nieuwenhuijsen et al. 2011) and the reduced muscle strength (Wiley and Damiano 1998), may be important factors for developing and sustaining physical fatigue in this group. Therefore, the deterioration of walking may be closely related to increased physical fatigue, which was investigated in Study I. Because of the known increased energy expenditure and physical fatigue levels in this group, a theoretical framework linking increased work during walking to increased fatigue, and subsequent walking deterioration was investigated in Study III.

Physical fatigue in CP might be partly understood within the frameworks of the “physiological burn-out syndrome” (Pimm 1992). This syndrome is assumed to be caused by an imbalance between physiological resources and the environmental demands over time. The symptoms are described as reduced muscle strength, loss or significant reduction of hand function, worsened ability to speak (if the person already has dysarthria) and physical fatigue (Pimm 1992). This was most often seen in individuals who had a wide difference between the demands of daily life and their physiological resources. Bottos and Gericke (2003), in their review on walking function in adults with CP, concluded that joint deterioration, physiological (or psychological) burn-out, fatigue and surgery (either too much or the decision not to have a particular intervention) were important factors in the loss of walking capacity in adults with CP.

The longitudinal development of fatigue in people with CP has only been studied by Malone and Vogtle (2009), who found relatively stable levels of fatigue in a group of 26 adults with CP, of which 14 were non-ambulatory. Whether fatigue levels increase or are stable over time in a group of mainly ambulatory adults with spastic CP are not known, and was therefore investigated in Study I.

2.5 HEALTH-RELATED QUALITY OF LIFE

HRQoL is commonly referred to as the extent to which health impacts on an individual's ability to function and his/her perceived well-being in physical, mental and social domains of life (Hays et al. 2002). HRQoL is the different dimensions related to limitations in health, and can be claimed to cover broad categories, such as duration of life, impairment, opportunities, functional status, and health perceptions (Patrick and Bergner 1990). HRQoL is a part of the broader construct of Quality of life, which includes aspects of life such as inter-personal relations, spiritual and aesthetical experiences. In the studies in this thesis the HRQoL was of particular interest, because of its relation to walking and functional mobility, as operationalised in SF-36 (Ware 1997).

2.5.1 Physical and mental components of HRQoL in adults with CP

In adults with CP, HRQoL was investigated in a few studies (Gaskin and Morris 2008, van der Slot et al. 2010, Jahnsen 2004). The two former studies found generally lower HRQoL compared to the GP, especially for physical functions, while the role limitations due to mental problems and mental health were not significantly lower than in the GP. Jahnsen et al. (2004) has found significantly lower levels for all HRQoL domains except for vitality, and Sandström et al. (2004) found reductions in all health domains, but most in the mobility and pain/discomfort domains. Van der Slot et al. (2010) found better scores on the physical and mental health scales (PCS and MCS) in those who scored higher on the effort subscale of self-efficacy, indicating that a greater willingness to expend effort in completing behaviour was associated with better PCS and MCS scores.

Physical and emotional role functions are parts of HRQoL, and reductions in these were associated with pain and fatigue in adults with CP (Jahnsen et al. 2004a, Jahnsen et al. 2003b). Both pain and fatigue were associated with deterioration of functional skills. Because pain seemed to be an important factor for walking deterioration, it was also important to understand more about pain and its relationship to physical and mental components of HRQoL, and this was investigated in Study II.

Health and well-being in adults with CP has been addressed in two recent review articles (Liptak 2008, Turk 2009). A common finding was that adults with CP decrease their utilisation of health care and related resources, and they have an increased prevalence of co-morbid and secondary conditions, such as pain, fatigue and mobility reductions. They have less participation in social interactions, employment, marriage and independent living than adults without CP (Liptak 2008). Despite this, Turk (2009) found that most adults with CP have good self-reported health, but also recognise the need for more studies of the interplay between health, HRQoL, wellness, disability and social participation.

The finding that many persons with a high degree of disability also report good and excellent quality of life, despite health problems and mobility problems, has been called the "disability paradox" (Albrecht and Devlieger 1999). Drum et al (2008) have found that people with disabilities reported overall lower self-reported health, and more days of physical and mental ill-health. However, within the same self-rated health category, the people with disabilities had more days of physical and mental ill-

health, suggesting that the tolerance of ill health is greater in individuals with disabilities, and despite fewer days with good health the self-rated health was still good. This suggests that the understanding of the concept of health might differ between people with and without disabilities (Drum et al. 2008).

2.6 BALANCE

The ability to control body position in relation to the task, the environment and the individual constraints is fundamental to almost all aspects of human functioning in everyday life, thus all tasks require postural control, containing an orientation and a stability component (Shumway-Cook and Woollacott 2007). Postural orientation is defined as: “the ability to maintain an appropriate relationship between the body segments and between the body and environment for a task” (Horak and Macpherson 1996). The postural stability is defined as: “the ability to control the centre of mass in relationship to the base of support” (Shumway-Cook and Woollacott 2007). In this thesis, balance includes both postural orientation and stability dimensions, and the term balance is thus used synonymously with postural control.

The “systems approach” to the understanding of motor control was first described by Bernstein (1967). A “systems approach” sees motor control as organised around behavioural goals, and the movement strategies emerge from a continuous interaction between the individual and the environment in order to fulfil the task. The movements are organised as an interaction between different systems in a heterarchical manner. All of these systems contribute with different aspects of control and none of them can be said to govern over the other systems (Shumway-Cook and Woollacott 2007). Therefore, the movement cannot be considered in isolation from the task, the environment and the context in which it takes place, and it is a dynamic interplay between perception, cognition and action systems (Shumway-Cook and Woollacott 2007). Thus, to be able to maintain balance, a complex process that involves the integration of information from different sensory systems, orientation of the body segments, and between the body and the environment for a task, are necessary (Horak and Macpherson 1996). This also includes information from vestibular, somato-sensory, visual and musculoskeletal systems, in the presence of cognition which includes attention and reaction time (Muir et al. 2010).

The subsystems that are important for successful balance control have been described by Horak (2009) as being biomechanical constraints, limits of stability/verticality, anticipatory postural adjustments, postural responses, sensory integration and stability in gait. The biomechanical constraints relate to the joint ROM and muscle strength, as well as the alignment of the body segments. The limits of stability/verticality refer to the sense of where the projection of the centre of mass is in relation to the base of support, as well as to the sense of gravitational direction and the midline of the body. The anticipatory postural adjustments ability, or the feed-forward control, refers to the preparatory movements or muscle actions that are made before a movement has started, in order to prevent the destabilising effect of the movement (Shumway-Cook and Woollacott 2007). Postural responses refer to the movements that occur in response to sensory feed-back. The feed-back can come from different sensory systems, such as visual, vestibular and somatosensory, and can be a response to unexpected slips or trips, as well as external perturbations when the surface moves, like for instance on an escalator (Shumway-Cook and Woollacott 2007). The sensory integration system allows for the integration, and interpretation of signals from the

different sensory systems, such as vision, the somato-sensory system and the vestibular system (Horak 2006). Stability in gait refers to the dynamic control of balance during walking, and the dynamic coordination between the motor system and the somato-sensory systems. The role of cognition is also a part of this system, as cognitive tasks require cortical attention, and thus compete with the other neural systems if the attentional and executive systems are constrained (Horak et al. 2009).

2.6.1 Balance in individuals with CP

For people with CP, reduced balance has been described as one of the primary abnormalities of gait (Gage et al. 2009), and stability in the stance phase is referred to as the first prerequisite of gait, highlighting its importance for gait (Gage et al. 1996). Balance was found to be impaired in children with CP (Burtner et al. 2007, Liao et al. 1997, Rose et al. 2002). A proximal-to-distal muscle activation pattern (Nashner et al. 1983, Burtner et al. 1998), increased co-contraction (Brogren et al. 2001, Burtner et al. 1998), and slower muscle activation as a response to balance threats (Brogren et al. 2001) are key differences compared with typically developing children. Similar studies in adults with CP have not been carried out, but there is reason to assume that the same balance impairments are present.

In the study by Jahnsen et al. (2004b) reduced balance was the most frequently reported description of walking deterioration, reported by 63%, thus indicating that reduced balance might be an important factor for walking deterioration in spastic CP. Balance and its relationship with walking deterioration in adults with spastic CP has not been examined, nor has the balance been characterised in this group. This was therefore the focus of Study IV.

The subsystems described by Horak et al. (2009) offer a useful framework that may reflect how the impairments that are associated with CP could cause balance limitations. Biomechanical constraints may be present because of muscle weakness and reduced joint ROM. The limits of stability might be reduced because of sensory deficits. The anticipatory adjustments may be limited due to delayed motor learning and reduced motor control as well as increased muscle tone. Postural responses may be affected because of sensory deficits, as well as increased muscle tone, antagonist co-activation and reduced motor control. Sensory integration can be affected by sensory deficits in visual, somato-sensory or vestibular systems. Stability in gait can be reduced by reduced motor control, increased muscle tone, and deficits in coordinating motor systems for gait and somato-sensory systems dynamically (Horak et al. 2009). Simultaneous cognitive tasks during motor activities may interfere in the use of cortical attention, and lead to instability in gait. As adults with CP experience other problems, such as pain and fatigue, in addition to their neurological impairments, these problems may also affect their balance.

2.7 ICF

The International Classification of Functioning, Disability and Health (ICF) offer a useful framework to consider the impact of impairments associated with CP and consequences on activity and the participation restrictions, as well as environmental and personal factors, as all these factors have an impact on and are being affected by the health status. The World Health Organisation (WHO) published the ICF in 2001 (WHO 2001). ICF allows for the classification of impairment, function and disability

in different domains. These domains are the body functions and structures, as well as activity and participation. Further, the environmental and personal factors may have both positive and/or negative influences, and all of these domains may impact or being impacted by the health status (Figure 3). The ICF is not a hierarchical structure, but the underlying understanding is that the different domains can affect one another in all directions, thus reflecting a bio-psycho-social model of health (Figure 3).

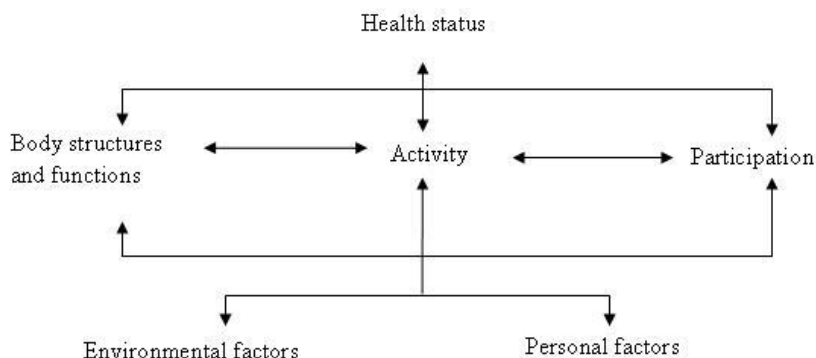


Figure 3. ICF model of interaction between different life areas and health.

Core sets of ICF categories to include in assessments of patients have been developed for a range of chronic conditions (Cieza et al. 2004), but not for adults with CP. The different aspects of a disability such as CP can be classified according to the ICF, but there is no unified consensus about which categories to include in the different domains. There are several different classification systems in use today that may be sorted according to the domains of body structures and functions, as well as in activities, but there is no standardised scheme to describe participation restrictions in CP (Rethlefsen et al. 2010). The different descriptive and outcome variables that are used in this thesis are sorted according to ICF domains, and are presented in Table III.

3 AIMS OF THE STUDY

The general aim was:

To investigate walking over a seven-year period, to characterise and investigate the critical variables associated with walking deterioration and the changes in these variables over seven years, and to compare and characterise gait and balance in relation to walking deterioration in adults with spastic cerebral palsy.

Specific aims were:

To compare pain, fatigue, walking deterioration and gross motor function over a seven-year period in a group of individuals with spastic unilateral and bilateral CP, and to investigate the relationship between walking deterioration and pain, fatigue and gross motor function (Study I)

To characterize the prevalence of pain at different body sites, compare the prevalence over a seven-year period, and to investigate the relationship between number of pain sites and physical and mental components of Health Related Quality of Life (Study II)

To compare work of walking and kinematic gait variables between adults with spastic bilateral CP reporting walking deterioration and those who do not, and to characterise kinematic and kinetic gait variables across all participants (Study III)

To compare balance confidence, fear of falling and balance ability between adults with spastic bilateral CP reporting walking deterioration and those who do not, to characterise balance confidence, fear of falling and balance ability, and to investigate the relationship between balance confidence and balance ability across all participants (Study IV)

4 MATERIAL AND METHODS

4.1 STUDY PARTICIPANTS

The participants were recruited from a cross-sectional national rehabilitation-based survey of adults with CP in Norway in 1999 (Jahnsen 2004). A flowchart of the whole inclusion process with both data collections is presented in Figure 3.

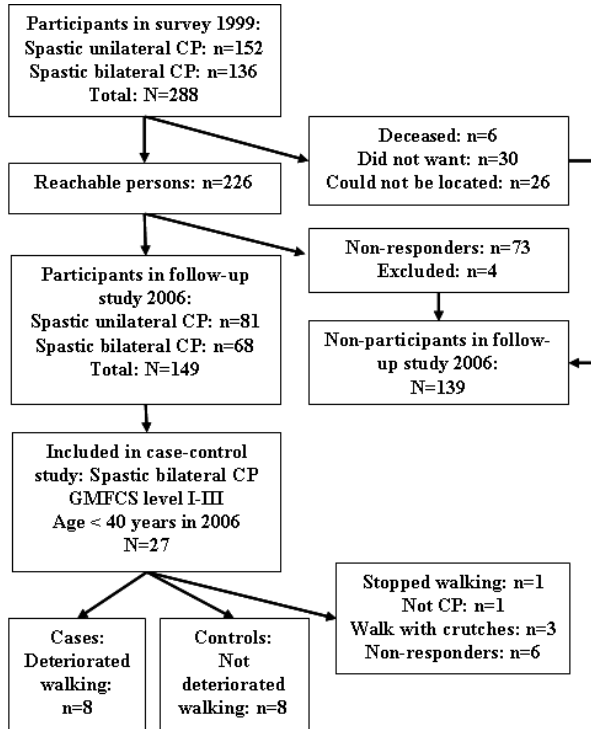


Figure 4. Flowchart of the inclusion of participants in the four studies in this thesis.

4.1.1 Participants in Studies I and II

Inclusion criteria were participants in the 1999 survey (Jahnsen 2004), unilateral and bilateral spastic CP, Gross Motor Function Classification System (GMFCS) levels I-IV in 1999, and having at least been able to walk indoors, with or without support. Exclusion criteria were individuals with dyskinetic, ataxic and bilateral CP with GMFCS levels IV-V in 1999 which had not been able to walk indoors with help, and documented intellectual disabilities. Diagnosis was self-reported, but checked against medical records when in doubt and when such records were available. Demographic and diagnostic information about the participants, together with the same information from all participants in the original study in 1999, is presented in Table I. A non-responder analysis was carried out, showing no differences between the participants and the non-responders. Co-morbidities other than CP were self-reported in 1999. Of the participants in the follow-up study in 2006, fifty-five reported other diseases than

CP in 1999. The most common were allergies (n=27), epilepsy (n=24) and other (n=12). Co-morbidities were not reassessed in 2006, but four individuals spontaneously reported additional diagnosis (multiple sclerosis, n=3 and rheumatoid arthritis, n=1). These were considered to potentially influence walking negatively, and were therefore excluded.

Table I. Demographic and diagnostic distribution of the present respondents compared with those in the original study by Jahnsen et al. (2004b).

			Study population [2006] (N=149)	Original study [1999] (N = 288)	
Sex:	male/female, n		76/73	153/135	
Age (years: months) (SD), min-max			40:5 (10.7), 24-76	33:6 (11.1), 18-70	
Height (cm), mean (SD)			169.9 (10.4)	169.7 (10.3)	
Body weight (kg), mean (SD)			73.0 (16.7)	69.0 (14.5)	
			%	%	
Diagnosis [2006]	GMFCS level	I	65	79	
Unilateral CP n=81		II	20	10	
		III	5	9	
		IV	7	3	
		V	3	0	
Bilateral CP n=68		I	22	25	
		II	31	18	
		III	24	46	
		IV	22	11	
		V	1	1	
Education:	First level (9 yrs)		16	25	
	Second level (12 yrs)		53	48	
	Third level (>12 yrs)		31	27	
Living situation:	Alone		45	43	
	With children		4	5	
	With parents		6	19	
	Married/cohabiting		43	28	
	With others		2	5	
	Source of income:	Paid job	100%	30	26
			50-99%	8	9
		<50%	5	4	
	Studying		4	10	
	Disability benefit				
		100%	36	38	
		50-99%	10	9	
		< 50%	0,6	0,03	
		Unknown	9	0	
		Others	23	13	
	Old-age pension		4	1	

4.1.2 Participants in Studies III and IV

The participants were recruited from a follow-up study of 149 adults with spastic unilateral and bilateral CP in 2006 (Opheim et al. 2009). In study I, walking deterioration was found to be most common in spastic bilateral CP, and in the 35-40 years age group. Therefore, all people with spastic bilateral CP, who reported GMFCS level I-III (Palisano et al. 1997) and were below 40 years of age in the 2006 survey, received a mailed letter, and were invited to participate. One exclusion criteria was the inability to walk at least 10 m without walking aids. This criterion was chosen because the kinetic measurements of gait require walking without aids in order to be valid. A flowchart of the inclusion process is presented in Figure 4. The diagnostic and clinical characteristics of the participants in Studies III and IV are presented in Table II. The cases were individuals who reported walking deterioration, while the controls did not report this. The most common self-reported causes of walking deterioration were increased stiffness in muscles and joints (n=4), and poor balance/fear of falling (n=3). Thirteen of 16 participants had fallen more than five times last year. There were no differences between the cases and the controls in the descriptive variables. Fourteen of the participants had had foot/ankle surgery, 10 had had hip surgery, and four had had knee surgery. Twelve subjects with spastic bilateral CP, GMFCS I-III, some recruited from the participants in Studies III and IV, and others inpatients from Sunnaas Rehabilitation Hospital participated in an intra-rater reliability study on balance confidence, balance ability and gait. This was conducted by the same principal investigator as in the present studies, with a median of one day between tests.

Table II. Characteristics of the participants in Studies III and IV (N=16).

	All	Cases (n=8)	Controls (n=8)
Females/males, n	11/5	5/3	6/2
Age, years, median (Q1-Q3)	38 (32-40)	39 (31-42)	36 (31-39)
Body Mass Index, median (Q1-Q3)	24.7 (23.1-31.4)	25.0 (23.5-37)	24.7 (22.7-27.1)
GMFCS II/III, n	15/1	8/0	7/1
Muscle tone ¹ (0-5), median (Q1-Q3)	2 (1.6-2.5)	2 (2-2.4)	2 (1.1-2.9)
Muscle strength (0-5), median (Q1-Q3)	4 (4-4)	4 (3-4)	4 (4-4)
Falls last month, n = 0	7	3	4
1-2	4	2	2
3-8	4	2	2
20	1	1	0
Falls last year, n <5	3	1	2
5-10	6	3	3
11-25	3	1	2
26-50	1	1	0
>51	3	2	1

¹Modified Ashworth scale, higher scores indicate a higher muscle tone and resistance to passive movements, and the scores were coded as: 0=0, 1=1, 1+=2, 2=3, 3=4, 4=5.

4.2 METHODS

The descriptive and outcome variables used in Studies I-IV are sorted according to the relevant ICF domains (Table III). A number of different variables were chosen to provide a multidimensional assessment, as described by Thomasson et al. (2011).

Table III. Descriptive and outcome variables in this thesis sorted according to the ICF domains. The studies in which they were used are specified and numbered I, II, III and IV.

Body structures	Function and impairment			Contextual factors
	Body functions	Activity	Participation	Environmental and personal factors
s750 Skeletal deformities (III&IV)	b280 Pain (I&II)	d4 General mobility (I)	p8 HRQoL (I&II)	e1 Walking aids, wheel chair (I)
s750 Joint ROM (III&IV)	b770 Gait analysis kinematics and kinetics (III)	d8 HRQoL (I&II)	d8 Education (I&II)	Mental components of HRQoL (II)
	b730 Muscle strength (III&IV)	d450 GMFCS Walking (I)	d850 Work (I&II)	Physical components of HRQoL (II)
	b735 Muscle tone (III&IV)	d4 Balance ability (IV)	Mental components of HRQoL (II)	
	b156, b164, b235 Balance confidence (IV)		Physical components of HRQoL (II)	
	b156, b164, b235 Fear of falling (IV)			
	b130, b455 Fatigue (I)			
	b156, b164, b760 Balance ability (IV)			
	b710 Joint ROM (III&IV)			

4.2.1 Methods in Studies I and II

Design: Seven year follow-up study

Questionnaire: The same questionnaire and the same methods as in 1999 were used, with the addition of the Fatigue Severity Scale (FSS) (Krupp et al. 1989). The questionnaire was sent by mail, with a prepaid return envelope. A written reminder was sent after approximately three weeks. The questionnaire contained questions on the following areas:

Walking was assessed with self-reported Gross Motor Function Classification System (GMFCS) level (Palisano et al. 1997). In 1999, GMFCS levels were assessed retrospectively, based on clinical and medical records information. In 2006, GMFCS levels were self-reported and checked with other reports on motor function and medical charts when in doubt and when these were available. The agreement between self-reported and professionally rated GMFCS levels was found to be excellent (Jahnsen et al. 2006).

Walking deterioration was assessed with the statement: My walking function has: become better over the years, neither better nor worse, or become worse over the years. Improvements in walking function were described as: improved balance, stronger muscles, improved cardio-vascular fitness, higher walking speed or others. Deterioration in walking was described as: worse balance, weaker muscles, worse cardio-vascular fitness, slower walking speed, or others. More than one alternative response was possible, as well as adding own comments. The age of either improvement or deterioration of walking function was given with check boxes with five-year intervals.

Pain was assessed using the same questionnaire as in 1999. Pain frequency, pain duration, pain sites and factors that improve (pain medication, activities, physiotherapy, rest or other) or worsen the pain (exhaustion, activities, rest, other) were registered. In Study I the subscale of bodily pain (BP) from SF-36 was used to assess the combination of pain intensity and the impact of pain on work and daily activities (see HRQoL below) (Ware 1997, Loge and Kaasa 1998).

Fatigue, or the experience of feeling exhausted, tired, weak, or lacking energy (Kaasa et al. 1999), was measured using two scales, the Fatigue Questionnaire (FQ) (Chalder et al. 1993), and the Fatigue Severity Scale (FSS) (Krupp et al. 1989). FQ was used in the original study, and has a GP-based norm, thus allowing for comparisons (Loge et al. 1998a). The age distribution in the reference material for FQ is the same as for SF-36. Chronic fatigue was considered according to Loge et al. (1998a), as a dichotomised score above 4, lasting for longer than six months. FSS was added because it does not require the respondents to compare fatigue status retrospectively, and explores the agreement with different expressions related to the impact of fatigue. FSS has been translated and validated in Norway, and has GP-based norms, and an FSS score above 5 is suggested to reflect high degree of fatigue (Lerdal et al. 2005).

HRQoL was assessed with the Medical Outcome Study Short Form 36 (SF-36) (Ware 1997, Ware and Sherbourne 1992). SF-36 has been used extensively in hundreds of studies throughout the world, with many different patient populations. The reliability and validity of SF-36 has been tested in 45 countries in the International Quality of Life Assessment Project (IQLA 2011, Ware and Gandek

1998, Aaronsen et al. 1992, Alonso et al. 2004). SF-36 has been translated into Norwegian, and has GP-based norms (Loge and Kaasa 1998, Loge et al. 1998b). SF-36 assesses HRQoL in eight different domains (Alonso et al. 2004, Ware, Jr. et al. 1998). These domains are: social function, physical function, mental health, general health, vitality, emotional role function, bodily pain, physical role function. The domains are scored on a 0-100 scale, where 0 is the worst and 100 is the best. In Study II, the eight SF-36 subscales were used to form aggregated composite scores of the mental (MCS), and physical component summaries (PCS). PCS and MCS were computed with standardised, weighted scores from the eight physical and mental subscales (Ware et al. 2000). MCS and PCS were standardised with a linear T score transformation to norm-based scores from the GP in Norway (Loge and Kaasa 1998, Ware et al. 2000). The norm-based scores have a mean of 50 points, and a standard deviation (SD) of 10 points. Scores higher than 50 are better than the GP mean. MCS has been found to be valid measures for mental health (Gill et al. 2007) and may therefore be used as an indicator of psychological health. PCS has been found to be associated with impairments in the lower extremities (Hall et al. 2000) and may be used to indicate physical impairments. Due to missing data on one or more of the SF-36 subscales, PCS and MCS were calculated from 131 persons.

4.2.2 Methods in Studies III and IV

Study design: Case-control design. Those reporting walking deterioration were considered cases, and those not doing so were controls.

The outcome measures in Studies III and IV were chosen mainly based on the findings from Studies I and II, and are described in the sections “Theoretical framework for between-group comparisons”, and “Preparations for Study IV” (see below). Three-dimensional gait analysis (3DGA) was chosen as the best method to study gait, as this gives detailed descriptions of lower extremity joint angles, joint moments and joint powers in three planes, and allows for the computation of work of walking and the categorisation of sagittal gait patterns.

Data collection procedure: The Activities-Specific Balance Confidence Scale (ABC) was administered in a telephone interview by a physiotherapist 2-3 weeks before the semi-structured interview and the tests of balance ability. This physiotherapist did not take part in the other assessments. In the semi-structured interview, conducted by the principal investigator (AO), the changes in walking function over the years (since the end of adolescence) were reassessed, and categorised as improved, unchanged or deteriorated. The spontaneously mentioned causes of deterioration were recorded. The Falls Efficacy Scale-International (FES-I), the Functional Mobility Scale (FMS), GMFCS and history of falls were also obtained during this interview. All assessments were carried out in the same order: first the semi-structured interview, then clinical examination, followed by the tests of balance ability and the gait analysis, performed on a single occasion. Balance ability tests were always carried out in the same order, first the Four Square Step Test and then the BESTest. The principal investigator led and administered both the clinical examination and all the balance tests, assisted by at least one other person (physiotherapist and/or biomechanist) who observed and registered the performance of the balance tests, and aided in securing the participants’ safety. When there were doubts over scoring, this was discussed, and the principal investigator had the final word. The principal investigator was blinded to the ABC scores until after the tests of

balance ability, in order not to be influenced by the balance confidence reported by the individual.

Descriptive variables study III and IV

Walking was described by the Functional Mobility Scale (FMS) (Graham et al. 2004) and the GMFCS (Palisano et al. 1997). To differentiate between GMFCS level I and II, the use of railing when ascending or descending stairs was used.

Passive range of motion (ROM) in hip, knee and ankle joints was measured using a goniometer (Nortkin and White 1985). The end ROM was measured when the resistance was initially felt, and not stretched to the individuals' tolerance (Gage et al. 2009)(p193). The variation between testers of joint ROM is found to be around 10° (McDowell et al. 2000).

Muscle tone of the triceps surae, rectus femoris, hamstring and adductor muscle groups was assessed bilaterally using the modified Ashworth scale (MAS) (0-5) (Bohannon and Smith 1987). This scale assesses resistance to rapid, passive movements. The median MAS score of all these muscles were calculated for each individual to form an index (Østensjø et al. 2004). Bohannon and Smith (1987) found agreement of 87% between two raters of MAS in elbow flexors, but the reliability of MAS has been found in later studies to be variable (Bhimani et al. 2011). The validity of MAS in the assessment of muscle spasticity has been questioned (Bhimani et al. 2011, Fleuren et al. 2010), but it is still the most common clinical scale for assessing muscle tone by grading the resistance to passive movements (Ahlborg et al. 2006), acknowledging that this also includes the mechanical compliance in muscle and joints (Patrick and Ada 2006).

Muscle strength in hip flexors, extensors and abductors, knee flexors and extensors and ankle plantar and dorsiflexors bilaterally was assessed using the 0-5 grade Manual Muscle Test scale (Hislop and Montgomery 2007). Interrater reliability of manual muscle test was found to be 90% and intrarater reliability was even higher, and for summary scores of muscle tests the average difference was 4% (Iddings et al. 1961). The manual muscle test was found to correlate ($r=0.768$, $p<0.001$) with dynamometer testing (Bohannon 2001). The median muscle strength score of all these muscles was calculated for each individual to give an overall indication of muscle strength (Lohmann Siegel et al. 2004).

Selective Motor Control (SMC) (0-4) of ankle dorsiflexors was assessed using the scale developed by Boyd and Graham (Boyd and Graham 1999). Higher scoring on the scale indicates a better ability to isolate the activity of the tibialis anterior muscle from the other muscles in the leg. The SMC was found to have moderate inter-rater reliability (Cohen's Kappa 0.55) for children with CP in the age of 5-7 (Smits et al. 2010), and is widely used in children with CP (Lofterod et al. 2011), but its reliability has not been reported in adults with CP.

Outcome measures in Study III

The 3DGA was carried out at the Motion Analysis Laboratory of Sunnaas Rehabilitation Hospital. Six infrared MX13 cameras (Vicon Motion Systems, Oxford, UK) and two AMTI OR6-7 force plates (AMTI, Watertown, USA) were used, and data was captured at a 100 Hz sampling rate. Retro-reflective markers were placed on the anatomical landmarks according to the Plug-in-Gait model (Vicon Motion Systems, Oxford, UK). An eight-segment model was used, consisting of two feet, two

shanks, two thighs, a pelvis and a thorax segment, allowing for the calculation of whole body centre of mass (COM). The participants walked on a 10 m walkway, and data was captured from a 4-5 metre central zone. The participants walked barefoot at self-selected, comfortable speed. At least five trials with clean force plate strikes, preferably with both feet hitting the force plates consecutively were captured. The data was processed using the Nexus 1.4 software (Vicon Motion Systems, Oxford, UK). Marker trajectories were filtered with a spline filter with a mean square error (MSE) value of 10. Kinetic (moments and powers) and kinematic (joint angles) variables were computed with the commonly used Plug-in-Gait software (Vicon, Oxford, UK), normalised to 0-100% of a gait cycle and presented in graphs. One gait cycle on the right and left side from each trial, and a total of five trials from each participant were used in the analysis. The data processing was done by one independent person, and checked by the principal investigator.

The intra-rater reliability study (n=12) showed that the standard error of measurement (SEM) and the minimal detectable change (MDC) for a number of kinematic variables were in the range of 2-4° and 5-9°, respectively. McGinley et al. (2009) have in a systematic review of the reliability of 3DGA suggested that an error of measurement of 5° were within acceptable limits.

Theoretical framework for between-group comparisons

Three factors were identified, each consisting of variables chosen from biomechanical principles underlying gait analysis and considered as potentially associated with walking deterioration. In Factors I and II, it was hypothesised that increased work of walking (i.e. energy requirements) was associated with walking deterioration. Fatigue was assumed as a possible intermediate element between the two concepts, as significantly higher fatigue was found in adults with spastic CP reporting walking deterioration (Opheim et al. 2009), and higher fatigue scores were found in ambulatory than in non-ambulatory adults with CP, suggesting that fatigue was associated with strained walking (Malone and Vogtle 2009). In Factor III, it was hypothesised that larger gait deviations were associated with walking deterioration. Baker et al. (2009) found greater gait deviations in those with a higher GMFCS level.

Factor I Energy Requirements

Change in body centre of mass (COM) vertical height during walking was found to correlate strongly with energy expenditure in healthy individuals ($r=0.90$) (Kerrigan et al. 1995), and moderately ($r^2=0.60$) in individuals with myelomeningocele (McDowell et al. 2002). Furthermore, Bennet et al.(2005) found increased mechanical work from vertical COM displacements in children with diplegic CP. In this study, the work in changing the potential energy of COM (W_{COM}) during each step was calculated and expressed in J/kg/metre (Eq 1):

$$W_{COM} = ((\Delta COM_h * mg)/m)/step \text{ length} \quad (1)$$

As leg length can be considered proportional to step length, no further normalisation of ΔCOM_h was made. The median W_{COM} during one GC from five trials was used.

Minimal knee flexion in stance was also examined as an indicator of increased energy requirement during walking. It was additionally included in Factor I because a gait pattern with increased knee flexion in mid-stance was considered to be a very energy-demanding gait (Gage et al. 2009), but not necessarily including increased vertical COM displacement.

Factor II Joint Work

Mechanical joint work at the hip, knee and ankle joints was used to indicate energy requirements during gait. Increased work during walking was found to correlate with increased energy expenditure during gait in healthy individuals (Burdett et al. 1983). The total joint work (TJW), in J/kg, was calculated as the area under each joint power curve during one GC (Caldwell and Forrester 1992), by integrating the power by % GC and consisting of both positive and negative work.

$$TJW = \int_{t1}^{t2} |P_j| dt \quad (2)$$

The total limb work (TLW) was calculated as the sum of these joint works (Caldwell and Forrester 1992):

$$TLW = \sum \int_{t1}^{t2} |P_{jt}| dt \quad (3)$$

TJW and TLW were normalised in two alternate ways. Firstly, relative to stride length to obtain a measure of energy requirement pr metre walked (J/kg/m). Secondly, relative to walking speed to obtain a measure in J/kg/m*s⁻¹. The mean of the left and right leg from five trials, and the median values were used in the analysis.

Factor III Kinematic Gait Quality

The Movement Analysis Profile (MAP) and Gait Profile Score (GPS) (°) (Baker et al. 2009, Beynon et al. 2010) were used to quantify kinematic gait deviations. The MAP included the pelvis and hip in three planes, knee and ankle in the sagittal plane, and foot progression in the transverse plane. The MAP components were calculated as the root mean square (RMS) difference between the median of five curves from each participant, and that from a reference group of 50 healthy adults (mean age 39.7 SD 11.7 years) collected with an identical protocol at Sunnaas Rehabilitation Hospital. The GPS is the overall median of the individual RMS differences in the MAP.

Classification of sagittal gait patterns was determined by examination of each participant's sagittal kinematic curves by an independent physiotherapist with considerable experience in 3DGA and CP. The classification system by Rodda et al. (2004), based on the mid stance to toe-off part of the GC, was used to characterise participant's walking patterns as Group I: True equinus; II: Jump gait; III: Apparent equinus; IV: Crouch gait; or V: Asymmetric gait (Figure 5). Those within 1 SD of the reference group were classified as group 0: mild (Rodda et al. 2004). These broad patterns have been used for clinical management in children, but no application in adults has been reported.

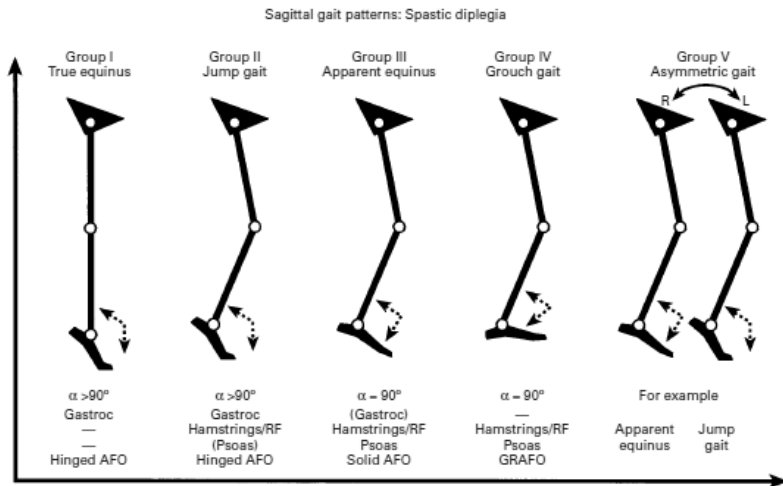


Fig. 1

Diagrams showing each gait pattern, with the dominant muscle groups identified for the management of spasticity and/or contracture and appropriate orthotic prescription. Group V is a combination of groups I to IV, with a different group in the right lower limb compared with the left lower limb. In this example, the right lower limb is group III, apparent equinus, and the left lower limb is group II, jump gait.

Figure 5. Gait patterns in spastic bilateral cerebral palsy. The dominant muscle groups identified for the management of spasticity and/or contracture and orthotic prescription [Reproduced and adapted with permission and copyright © of the British Editorial Society of Bone and Joint Surgery (Rodda JM, Graham HK, Carson L, Galea MP, Wolfe R. Sagittal gait patterns in spastic diplegia. *J Bone Joint Surg [Br]* 2004;86-B:251-8. Figure 1)]. AFO: ankle-foot orthosis; RF: rectus femoris; GRAFO: ground reaction force ankle-foot orthosis.

Preparations for Study IV

In order to gather more information about balance and the potentially appropriate outcome measures for a study on balance in adults with spastic bilateral CP, three adults were invited to share their experiences in a group discussion. They revealed that walking on uneven or slippery ground, walking in crowds, using escalators and public transport and walking down stairs were difficult situations that challenged their balance reactions. They often felt too slow and unable to prevent a fall. Impaired vision and reduced lighting conditions presented additional problems. These difficulties often led to compensatory strategies such as holding onto railings or other objects. Some regarded themselves as “expert fallers” i.e. they often fell but had not had serious injuries and were not afraid of falling. Others reported that they were afraid of falling, which restricted their participation in the community.

Outcome measures in Study IV

The following scales and tests of balance ability were chosen because they cover several of the balance problems mentioned in the group discussion, such as fear of falling, keeping balance while performing different activities, balance reactions and stepping over obstacles.

The Activity-specific Balance Confidence scale (ABC) was used to quantify, from zero to 100 (0=not at all), a person’s confidence in not losing balance while performing 16 activities of daily living (Powell and Myers 1995). The scale was found to be valid and reliable in healthy and frail elderly, as well as in patients with Parkinson’s (Mak and Pang 2009, Lord et al. 2010). The SEM and MDC of the ABC in the intra-rater reliability study were found to be 4.5 and 12.3 percentage points, respectively.

The Falls Efficacy Scale-international Norwegian version (FES-I) (Yardley et al. 2005, Helbostad et al. 2010, Helbostad and Yardley 2010) was used to measure fear of falling during different activities on a four-point scale (1=not at all concerned), giving a total scoring range from 16 to 64 points. It is widely used in the field of fall prevention, and found to be reliable and valid in community-dwelling elderly people, and elderly people treated for fall-related fractures (Delbaere et al. 2010, Nordell et al. 2009).

The Balance Evaluation System Test (BESTest) was used to describe different subsystems related to balance (Horak et al. 2009). The test contains six subscales covering six different subsystems of balance: I biomechanical constraints, II stability limits, III anticipatory postural adjustments, IV postural responses, V sensory orientation, and VI stability in gait. The BESTest consist of 27 items, some of which are divided into 2-4 sub-items (e.g for the left and right sides), leaving a total of 36 tasks (Table IV).

Table IV. Tests of balance and the subscales of the BESTest. The items are scored on a four-category ordinal scale (0=severe/unable, 3=normal).

BESTest I Biomechanical constraints (Scoring range 0-15)	1. Base of support (standing)
	2. Centre of Mass alignment
	3. Ankle strength and range
	4. Hip/trunk lateral strength
	5. Sit on floor and stand up
BESTest II Stability limits and verticality (Scoring range 0-21)	6. Sitting verticality and lateral lean (L and R)
	7. Functional reach forward
	8. Functional reach lateral (L and R)
BESTest III Anticipatory postural adjustments (Scoring range 0-18)	9. Sit to stand
	10. Rise to toes
	11. Stand on one leg (L and R)
	12. Alternate stair touching
BESTest IV Postural responses (Scoring range 0-18)	13. Standing arm raise
	14. In-place response-forward
	15. In-place response-backward
	16. Compensatory stepping correction-forward
	17. Compensatory stepping correction-backward
BESTest V Sensory orientation (Scoring range 0-15)	18. Compensatory stepping correction-lateral (L/R)
	19. Sensory integration for balance (mCTSIB)
	a) Eyes open, firm surface; b) Eyes closed, firm surface
	c) Eyes open, foam surface; d) Eyes closed, foam surface
BESTest VI Stability in gait (Scoring range 0-21)	20. Incline-eyes closed
	21. Gait-level surface
	22. Change in gait speed
	23. Walk with head turns-horizontal
	24. Walk with pivot turns
	25. Step over obstacles
	26. Timed "Get Up & Go" test
	27. Timed "Get Up & Go" test dual task

L: left, R: right; mCTSIB: Modified Clinical Test of Sensory Interaction on Balance.

All tests were scored on a four-category ordinal scale from 0=not able or absent to 3=normal. The scores were summed and reported as the percentage of maximum score on each subscale and total. The intra-rater reliability study showed the SEM and the MDC for the total BESTest score in percentage of maximum to be 1.6 and 4.4 percentage points, respectively.

Four Square Step Test (FSST) (Dite and Temple 2002) was used to test the ability to step over a low obstacle in all four directions in a given sequence as fast as possible, with shorter time in seconds indicating better performance. FSST was found to have a good test-retest reliability, correlated well with other gait measures (Whitney et al. 2007), and was feasible and valid for testing dynamic balance in ambulatory patients after stroke (Blennerhassett and Jayalath 2008). The intra-rater reliability study showed the SEM and MDC for the FSST to be 2.7 and 7.5 seconds, respectively.

4.2.3 Statistical methods

Descriptive statistics were presented with mean and standard deviation Studies I and II, and median and range or inter quartile range (Q1-Q3) or (min-max) in Studies III and IV. The analyses were made using the SPSS 14.0 and 15.0. In Studies I and II, changes in the period 1999-2006 were tested with the paired samples t-test for continuous and normally distributed data, and with the Wilcoxon sign rank test for ordinal and non-normally distributed data. As this was a follow-up study, a power analysis was not done, as we wanted to compare the respondents with themselves. Between-group differences in 2006 were tested using the independent samples t-test, for normally distributed data, and the Mann-Whitney U-test for non-normal or ordinal data (Altman 1991). Correlations between variables were tested with the Pearson's (Pearson's r) or the Spearman's correlation coefficient (Spearman's ρ). Correlations were interpreted according to Domholt: 0.00 – 0.25: little, if any; 0.26 - 0.49: low; 0.50 – 0.69: moderate; 0.70 – 0.89: high; 0.90 – 1.00: very high correlation (Domholt 2005)(p 358).

After the analyses for Studies I and II, the inclusion for Studies III and IV was carried out. There were 27 eligible individuals for these studies. A power analysis was carried out on the FSST, with an estimated standard deviation of 10 sec. This showed that with 10 individuals in each group, 90% power and a significance level of 0.05, a difference of 15.4 sec between the groups could be detected.

Due to the explorative nature of Studies III and IV, a conservative approach was adopted in the analysis, and between-group differences were tested using the Mann-Whitney U-test for both continuous and ordinal data. Relationships between variables were tested using Spearman's ρ .

For twelve subjects with spastic bilateral CP, GMFCS I-III, both the gait analysis and the balance tests were carried out twice, allowing for intra-rater reliability testing, which will be published elsewhere. However, we report here the standard error of measurement (SEM) [$SEM = \sqrt{\text{mean variance between tests}}$], and the minimal detectable difference (MDC) [$MDC = SEM * 1.96 * \sqrt{2}$], for the respective outcome variables in the methods section (Beckerman et al. 2001, de Vet et al. 2006).

4.2.4 Ethical aspects and protection of privacy in research

Approval for the study was given by the Regional Ethics Committee for Research in South-eastern Norway (ID: 1.2006.952). The Commissioner for the Protection of Privacy in Research (*Personvernombud*) gave approval of the information letter, consent letter and data storage. All participants gave written and informed consent.

5 RESULTS

5.1 STUDY I

The main findings in Study I was a significant increase in the number of individuals reporting walking deterioration among those with spastic bilateral CP, especially in the 35-40 years age group (Figure 6).

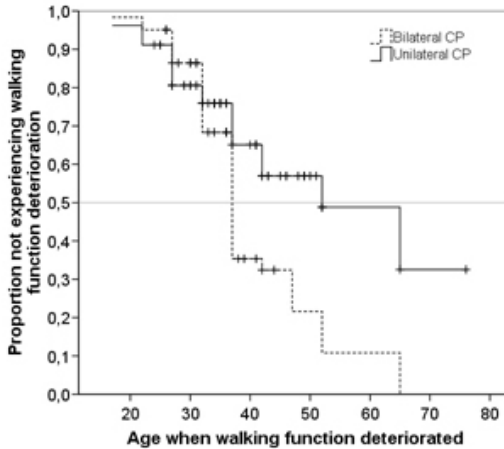


Figure 6. Kaplan Meier plot of the proportion of individuals not experiencing walking deterioration in adults with unilateral and bilateral CP. Vertical tic marks indicate where data have been censored, meaning that the individual have reached the age before a deterioration of walking has occurred [Published with permission from Wiley 2009. Opheim et al. 2009, *Dev Med Child Neurol*. MacKeithPress].

GMFCS levels in 1999 and 2006 are presented in Table V. Twenty-nine percent of those with unilateral CP, and 44% in bilateral CP had changed GMFCS levels. There were shifts between several levels in both directions, but especially between levels I and II. There were fewer shifts between other levels, except in bilateral CP, where nine out of 26 reported a reduction from level III to level IV.

Table V. Gross Motor Function Classification System (GMFCS) levels in 1999 compared with 2006 in a group of adults with unilateral and bilateral spastic CP in Norway (N=149). Higher GMFCS levels indicate gradually reduced Gross Motor Function. Grey cells on the diagonals show the number of individuals with the same GMFCS level in both 1999 and 2006.

			GMFCS 1999					Total
			level I	Level II	level III	level IV	level V	
Unilateral CP (n = 81)	GMFCS 2006	level I	48	5 ⁽⁺⁾	-	-	-	53
		level II	11 ⁽⁻⁾	4	1 ⁽⁺⁾	-	-	16
		level III	2 ⁽⁻⁾	-	3	-	-	5
		level IV	-	1 ⁽⁻⁾	1 ⁽⁻⁾	3	-	5
		level V	-	-	-	2 ⁽⁻⁾	-	2
	Total unilateral CP		61	10	5	5	-	
Bilateral CP (n = 68)	GMFCS 2006	level I	8	7 ⁽⁺⁾	-	-	-	15
		level II	9 ⁽⁻⁾	9	1 ⁽⁺⁾	1 ⁽⁺⁺⁾	-	20
		level III	-	2 ⁽⁻⁾	16	-	-	18
		level IV	-	-	9 ⁽⁻⁾	5	-	14
		level V	-	-	-	1 ⁽⁻⁾	-	1
	Total bilateral CP		17	18	26	7	-	

⁽⁺⁾⁽⁺⁺⁾ Indicates an improvement in Gross Motor Function corresponding to a shift of one⁽⁺⁾, or two⁽⁺⁺⁾ GMFCS level(s). ⁽⁻⁾⁽⁻⁾ Indicates a deterioration in Gross Motor Function corresponding to a shift of one⁽⁻⁾, or two⁽⁻⁾ GMFCS level(s).

From 1999 to 2006, there were no changes in the number of pain sites, pain intensity or the prevalence of chronic pain, but there was a significantly increased pain frequency from “seldom” to “monthly” ($p=0.002$), and significantly worse BP ($p=0.001$), indicating both higher pain intensity and a larger impact of pain on daily life and activities. From 1999 to 2006, there were no changes in physical, mental or total fatigue scores. When comparing those reporting walking deterioration versus those who did not, there was a higher prevalence of deteriorated walking with increasing GMFCS level ($p<0.001$) (Table VI). Further, those reporting deteriorated walking had significantly higher scores on pain frequency, duration, intensity and impact of pain on daily life and activities, as well as on physical fatigue and FSS (Table VI).

Table VI. Perceived changes in walking function over the years up to 2006 in adults with unilateral and bilateral spastic CP, in relation to diagnosis and gross motor function in the upper part of the table. The relation between perceived changes in walking and pain and fatigue scores is presented in the lower part of the table.

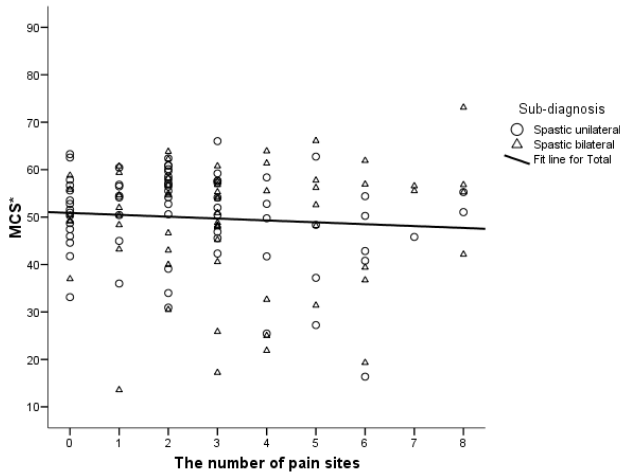
Perceived changes in walking up to 2006 (N=149)						
	Improved/ Unchanged (n=71) (48%)		Deteriorated (n=78) (52%)		Total (N=149)	p-value [95 % CI]
	n		n		N	
Diagnosis:						
Unilateral CP	51		30		81	<0.001
Bilateral CP	20		48		68	
GMFCS level						
I	50		18		68	<0.001
II	13		23		36	
III	5		18		23	
IV	3		16		19	
V	0		3		3	
	n	Median (Q1-Q3)	N	Median (Q1-Q3)	Median (Q1-Q3)	
Pain frequency, (1-5) ^a	64	2 (2-5)	76	4 (2-5)	3 (2-5)	0.014
No of pain sites, (0-9) ^b	54	2 (2-3)	69	3 (2-6)	3 (2-6)	<0.001
Pain VAS mm), mean (SD)	46	39.9 (22.8)	65	54.9 (23.2)	48.7 (24.1)	0.001 [-6.3 , -23.8] ^g
Bodily pain, (0-100) ^c	71	72 (51-100)	78	42 (32-62)	61 (41-84)	<0.001
FQ physical, (0-21)	70	7 (7-10)	77	10 (7-12)	8 (7-11)	<0.001
FQ mental, (0-12)		4 (4-4)		4 (4-5)	4 (4-5)	0.616
FQ total, (0-33)		11 (10-15)		13 (11-17)	12 (11-16)	0.001
FSS (1-7), mean (SD)	69	4.0 (1.4)	76	4.7 (1.4)	4.4 (1.4)	0.004 [-1.1 , -0.2] ^g

^aPain frequency registered as: 1: never, 2: seldom, 3: monthly, 4: weekly, 5: daily, 6: other. Category 6 is omitted from the statistics. ^bPain sites registered as: head, neck, shoulder, arms, back, hips, legs, feet and other. ^cSF-36 domain of bodily pain, higher scores indicate less pain intensity and impact of pain on daily activities. ^g95 % Confidence Interval of the mean difference between the improved/unchanged and deteriorated groups.

5.2 STUDY II

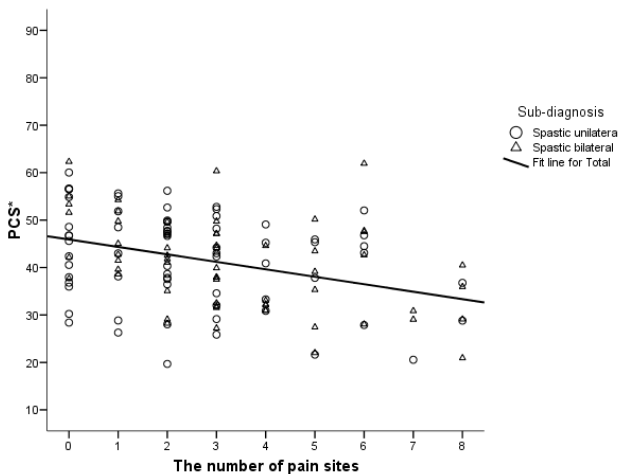
The main findings were little, if any correlation between the number of pain sites and mental components of HRQoL (MCS) (Pearson's r : -0.08, $p=0.38$) (figure 7a), while for the physical components of HRQoL (PCS), there was a low negative correlation with number of pain sites (Pearson's r : -0.36, $p<0.001$) (Figure 7b).

a



*Reference values for MCS are 50 points (SD 10). Lower score indicate worse score on the mental components of SF-36.

b



*Reference values for PCS are 50 points (SD 10). Lower score indicate worse score on the physical components of SF-36.

Figure 7a-b) Mental (MCS) and Physical component summary scale (PCS) of Health-related quality of life in relation to number of pain sites in a group of adults with spastic unilateral and bilateral cerebral palsy (N=131). The regression line for MCS (figure 7a) and PCS (figure 7b) is drawn. MCS and PCS norm-based score are on the Y-axis, with the mean norm score in the general population being 50 points with a standard deviation (SD) of 10 points. [Figures 7a-b reprinted from Opheim et al. 2011 J Rehabil Med (Study II), with kind permission from Journal of Rehabilitation Medicine].

PCS had a normal distribution, and a one-sample *t*-test showed that mean PCS (41.5 points SD 9.7) was significantly lower than in the GP, (mean difference -8.5 points [95% CI: -10.2 - -6.9] $p<0.001$). Median MCS was 53 points (Q1-Q3 45 - 57). MCS had more individuals reporting 1 SD above, and 2 and 3 SD below the mean in the GP (Z : 1.48, $p<0.025$), thus being statistically different from the GP. The prevalence rate of person reporting pain at the different sites, and the changes in prevalence from 1999 to 2006 are presented in Table VII.

Table VII. The prevalence rate (percentage) of persons reporting pain at different sites in 1999/2006 in adults with spastic uni- and bilateral CP (N=149). Significant changes from 1999 to 2006 are marked with asterisks: *($p<0.05$) or **($p<0.01$). Significant differences in prevalence rate between unilateral and bilateral CP, and between females and males in 2006 are marked with †($p<0.05$) or ††($p<0.01$), and the mark is placed at the group with the highest prevalence rate.

Pain site:	Unilateral CP (n=81)	Bilateral CP (n=68)	Females (n=73)	Males (n=76)	All (N=149)
Head	14/17	21/19	26/23	8/13	17/18
Neck	30/37	35/49*	44/52†	21/33*	32/42*
Shoulders	30/31	37/44	42/49††	24/25	33/37
Arms	25/25	24/21	34/30†	14/16	24/23
Back	36/52**	60/60	59/67††	36/45	47/56
Hips	25/27	32/43†	33/47* ††	24/22	28/34
Knees	28/19	37/31	36/33†	29/16*	32/24*
Foot/ankle	32/33	47/51†	49/51†	29/33	39/42

Back, neck and foot/ankle were the most common pain sites in both unilateral and bilateral CP. In unilateral CP there was a significant increase in back pain ($p<0.01$), and in bilateral CP, there was a significant increase in neck pain ($p<0.05$). Overall there was a reduction in knee pain ($p<0.05$). There was significantly higher prevalence of both hip and foot/ankle pain in bilateral CP ($p<0.05$). Females reported higher pain prevalence for all pain sites except for headache.

5.3 STUDY III

The main findings were the large deviations in kinematic and kinetic gait variables in the whole group. However, the between-group comparisons of work of walking and kinematic gait variables showed the following:

Factor I Energy Requirements: There were no differences between cases and the controls in the work of vertical COM displacement, ($p=0.645$) or minimal knee flexion in stance, $p=0.645$ and 0.195 for left and right leg, respectively.

Factor II Joint Work: There were no between-group differences in joint work at the ankle, knee and hip or total leg work normalised to distance or walking speed. There were considerable within-group variations in both groups, equally distributed within the groups. There was no difference in median (min-max) walking speed between cases, 1.07 m/s ($0.86 - 1.35$) and controls, 1.02 m/s ($0.83 - 1.31$) ($p=0.345$).

Factor III Kinematic Gait Quality: There was no difference in the overall GPS between cases, median 11.1° and controls median 12.7° ($p=0.141$) (Figure 8).

The overall MAP and GPS are presented in the lower panel of Figure 8. The largest deviations from the reference group occurred in the pelvic tilt, hip flexion/extension, knee flexion/extension and foot progression. The overall GPS was 11.7° (Q1-Q3: $9.1-14.3^\circ$).

The kinematic and kinetic curves are presented in Figure 9a-b (dark grey band), against reference group data (light grey band). Key abnormalities included increased anterior pelvic tilt, increased hip flexion during stance and terminal swing, and increased knee flexion at initial contact, loading response, mid-stance and terminal swing. The ankle showed a tendency of increased dorsiflexion in stance and reduced plantar flexion in terminal stance and pre-swing. A reduced knee extensor moment in stance was found.

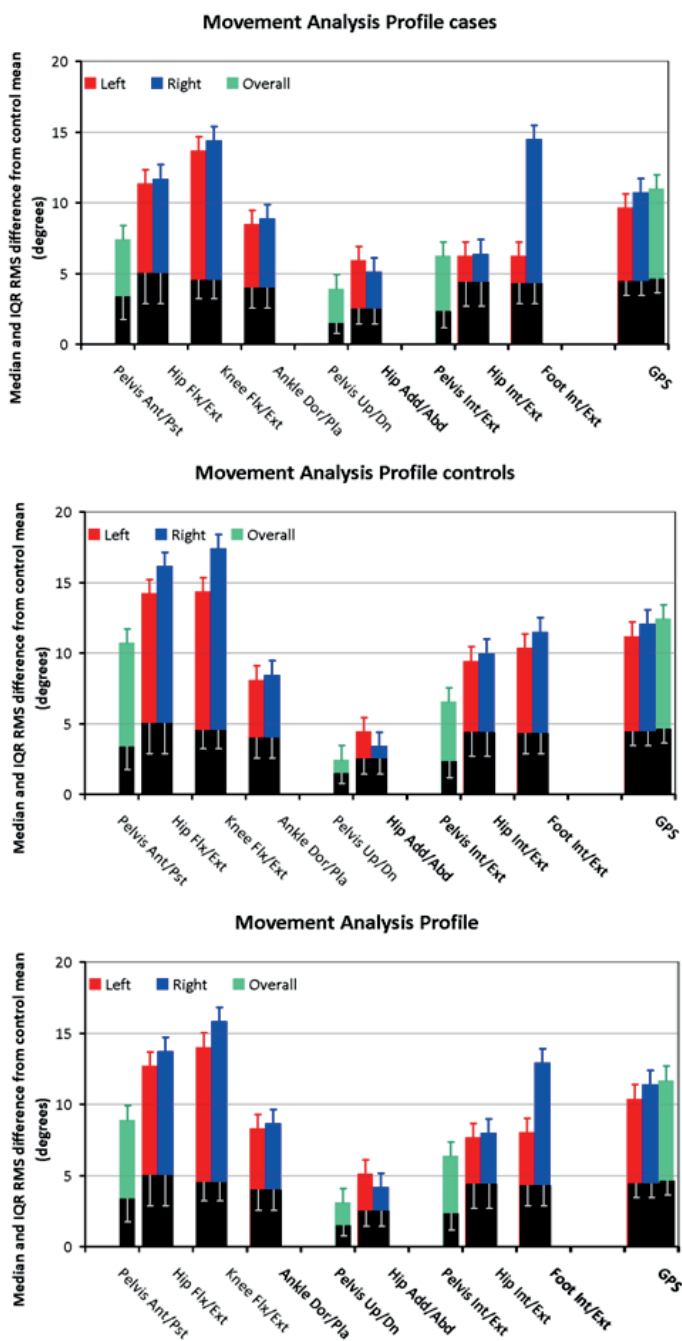


Figure 8. Movement Analysis Profile (MAP) and Gait Profile Score (GPS) in a group of 16 adults with spastic bilateral CP. The upper panel represents cases, reporting walking deterioration, the middle panel represents controls, not reporting walking deterioration, and the lower panel represents all participants.

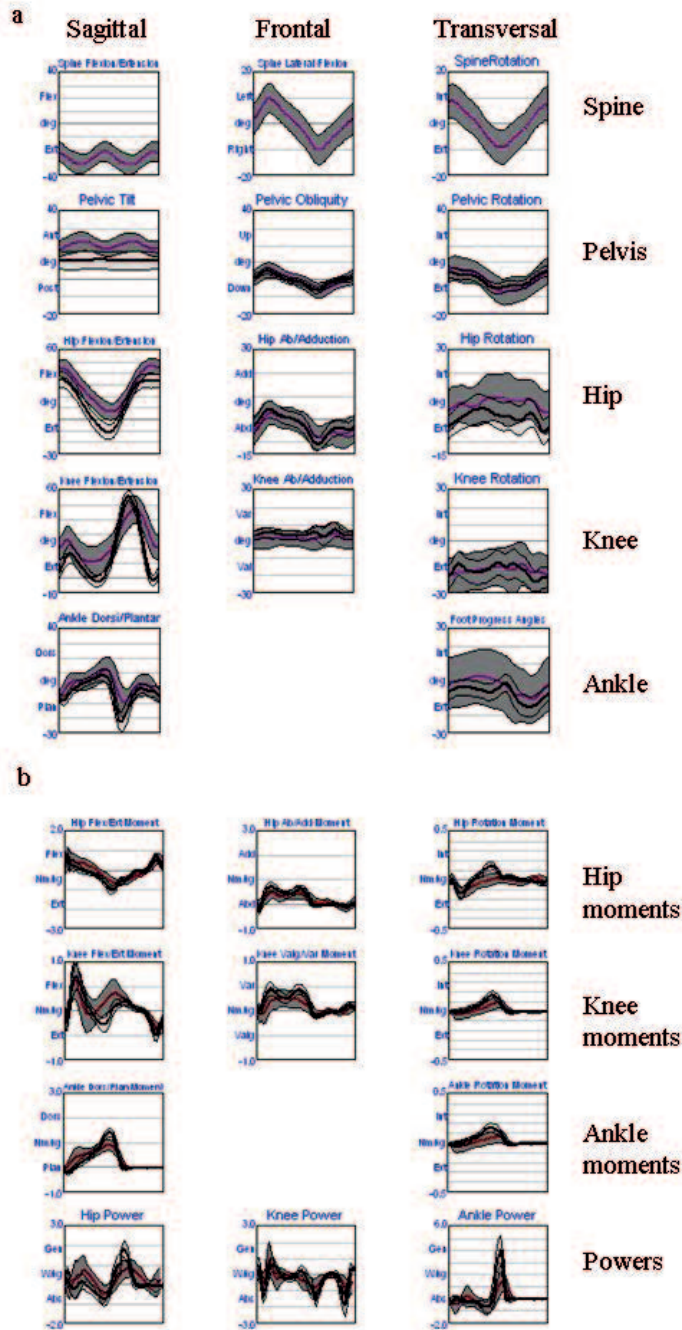


Figure 9a-b. Kinematic (panel a) and kinetic gait variables (panel b) in 16 adults with spastic bilateral CP. CP group, dark grey band, reference group light grey band with mean values and 1 standard deviation (SD), respectively. Panel a: Horizontal axis is 0-100% of gait cycle. Vertical axis is joint angle. Panel b: The fourth row is from left to right: hip, knee and ankle joint powers. All moments are external moments. Horizontal axis is 0-100% of gait cycle. Vertical axis is moment and power. Power above the zero line (positive values) is power generation, and power below the zero line is power absorption.

The classification of the sagittal gait patterns showed a majority of participants within group IV (crouch) (n=5) and V (asymmetric gait) (n=4). Group V contained two persons with the combination of apparent equinus and crouch, and one in each of the combinations crouch/mild and crouch/equinus. Fifteen of 32 limbs were classified as crouch and 8/32 as apparent equinus. Four persons (5 limbs) could not be classified according to the defined criteria. For example, in four limbs, the ankle and knee patterns lay within the reference band, while hip flexion and anterior pelvic tilt were excessive. Hence, the limb fitted neither the criteria for group 0 nor for group I. This was the first time this classification had been used in the adult CP population, and showed that the energy- demanding crouch gait pattern was common in this group.

5.4 STUDY IV

The main findings were the overall reduced balance ability, especially within the postural responses and anticipatory adjustments in the whole group. However, there were no differences in the balance confidence and fear of falling or balance ability between those reporting walking deterioration and those not (Table VIII).

Table VIII. Comparison of balance confidence, fear of falling and balance ability between people who reported deteriorated walking (n=8) and those who did not (n=8) in a group of adults with spastic bilateral CP (N=16). Medians and inter-quartile range (Q1-Q3). Scores on the subscales and total BESTest were converted to percent of maximum score (0-100%).

	Deteriorated walking	Non-deteriorated walking	<i>p</i> -value ^a	All
ABC scale ^b (%)	62 (32-76)	59 (46-70)	0.713	62 (37-70)
FES-I ^c (16-64)	24 (22-36)	26 (19-34)	0.804	24 (21-34)
BESTest I ^d (%)	57 (49-73)	60 (55-67)	0.798	60 (53-72)
BESTest II ^e (%)	76 (76-85)	84 (71-86)	0.645	79 (72-86)
BESTest III ^f (%)	39 (35-50)	44 (39-60)	0.328	42 (39-54)
BESTest IV ^g (%)	28 (18-63)	36 (18-55)	0.878	31 (18-58)
BESTest V ^h (%)	64 (45-78)	67 (50-73)	0.959	67 (50-73)
BESTest VI ⁱ (%)	67 (60-71)	69 (67-76)	0.505	67 (67-75)
BESTest total (%)	55 (40-67)	60 (57-71)	0.505	60 (55-68)
FSST ^j (s)	19 (15-28)	12 (10-20)	0.130	16 (11-22)

For the whole group, the ABC scale showed the lowest confidence in balance during the activities “stepping on or off an escalator without holding the railings”, “walking on an icy surface”, and “standing on chair and reaching” (Figure 10). Several of the items showed a ceiling effect with many attaining a very high score, for instance “walking around the house”, “sweeping the floor” and “walking to nearby car”. The overall ABC score across all activities was 62% (Q1-Q3: 37-70) confidence in not losing balance.

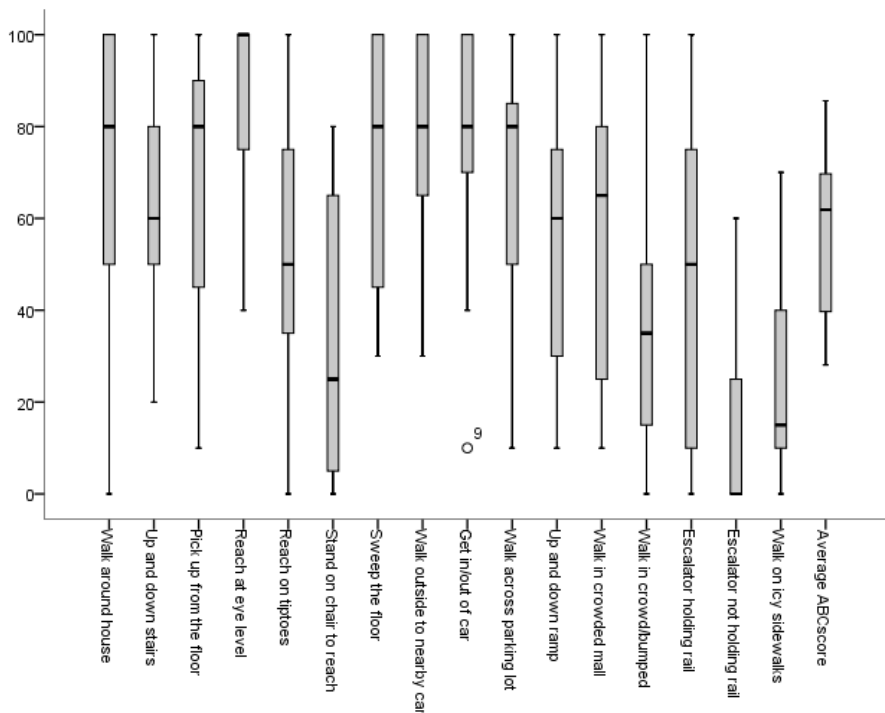


Figure 10. Boxplot of the scores on the different items of the Activities-Specific Balance Confidence (ABC) scale of 16 adults with spastic bilateral CP. 100 is 100% confidence in not losing balance during an activity, thus higher scores represent higher balance confidence. The boxes represent 50% of the observations (Q1-Q3), the black line shows the median score, and the error bars show the minimum and maximum values. The outliers are those that lie between 1.5-3 box lengths from the lower or upper edges of the box. [Reprinted from Opheim A et al. Balance in relation to walking deterioration in adults with spastic bilateral cerebral palsy. Accepted for publication in Physical Therapy. Reprinted with the permission of the American Physical Therapy Association. This is not the final, edited version.]

The FES-I showed the greatest fear of falling during the activities “walking on a slippery surface” and “walking up or down a slope”. The median summed FES-I score was 24 points (Q1-Q3: 21-34). In 12 out of the 16 items, the participants reported no or little fear of falling (Table VIII).

The BESTest showed the lowest scores on the anticipatory postural adjustments (Subscale III), median score of 42% (Q1-Q3: 39-54) and the postural responses (Subscale IV), with 31% (Q1-Q3: 18-58) of the maximum score. The median scores for the other subscales ranged from 60 to 79%, and the total BESTest score was 60% (Q1-Q3: 55-68) of the maximum score (Table VIII). The FSST showed a skewed distribution with two outliers, and the median time was 16 s (Q1-Q3: 11-22) (Table VIII). The relationship between balance confidence and balance ability showed a moderate positive correlation between the ABC scale and the BESTest subscales IV, V and VI, and total (Table IX).

Table IX. Correlations between balance confidence and balance ability in adults with spastic bilateral cerebral palsy (N=16). Correlation coefficients with 95% confidence intervals (CI) were obtained using Spearmans Rho (r_s); the CIs are given were significant correlations were found, $p<0.05$.

	Activities-Specific Balance Confidence Scale
BESTest ^a I Biomechanical constraints	0.45, $p = 0.19$
BESTest II Stability limits/verticality	0.24, $p = 0.36$
BESTest III Anticipatory postural adjustments	0.29, $p = 0.27$
BESTest IV Postural responses	0.52 [95% CI: 0.04, 0.81], $p = 0.04$
BESTest V Sensory orientation	0.52 [95% CI: 0.04, 0.81], $p = 0.038$
BESTest VI Stability in gait	0.57 [95% CI: 0.10, 0.83], $p = 0.022$
BESTest total	0.54 [95% CI: 0.06, 0.82], $p = 0.017$
Four Square Step Test	-0.346, $p = 0.189$

The tests of balance ability were easily understood by the participants, there were no major practical problems during the testing, and all participants completed all tests. Balance confidence, fear of falling and balance ability had not previously been characterised in this group before, nor had the correlations between balance confidence and balance ability been shown. These findings highlight the specific balance difficulties that this group experiences in daily life, but they could not explain the experience of walking deterioration.

6 DISCUSSION

The timing, extent, and localization of the primary brain lesion, result in a wide variety and combinations of clinical appearances of CP (du Plessis 2009). Further, the lesion affects motor function and sensory-motor development, postural control, as well as musculoskeletal growth. In adults with CP, the development of the impairments of body functions and structures takes place over a long time, and may have further varieties of appearances. In addition, both the conscious and unconscious adjustments that a person makes to cope with the impairments in daily life activities are highly individual. Thus, both the clinical appearances and coping with spastic CP are multi-faceted. This may be part of the reason why this is both a difficult and a scarcely researched area. Further, it may be difficult to have clearly defined and homogenous groups.

The secondary problems and complications in adults with spastic CP have been characterised as “(...) largely under-recognised, undocumented and under-treated”, and a higher focus on ambulatory functions was recommended as one of the research priorities by CPIRF (Cerebral Palsy International Research Foundation 2011). A gradual worsening of walking was described in childhood by both Johnson et al. (1997) and Bell et al. (2002) in adolescents up to 14 years of age. A gradual decrease of gross motor function in GMFCS III-V up to 21 years of age was reported by Hanna et al. (2009), and Jahnsen et al. (2004b) showed that at a mean age of 34, walking deterioration was reported by 44% and was increasing with age. The present studies show further deterioration of walking. The association between walking deterioration and gait and balance has been investigated and characterised in this group for the first time.

6.1 SEVEN-YEAR FOLLOW-UP

6.1.1 Walking

The main finding was that adults with CP experience a continuous deterioration in walking. The prevalence of this problem increased from 39% in 1999 (Jahnsen et al. 2004b) to 52% in 2006, and the problem was most common among individuals with spastic bilateral CP in the 35-40 years age groups. Some of the critical variables associated with walking deterioration have been identified previously (Jahnsen et al. 2004b, Murphy et al. 1995), and was followed-up in this study. Walking deterioration was associated with increased pain frequency, pain intensity, more pain sites, a higher impact of pain on daily life and physical fatigue. Andersson and Mattsson (2001) have found a somewhat lower prevalence of walking deterioration, 35%, in a CP population that was four years younger than those in this study.

Study I indicates that individuals with bilateral CP with GMFCS level III are more susceptible to walking deterioration than those with bilateral CP and milder impairments, and also compared to those with unilateral CP. One reason for this might be overuse (Pimm 1992), as the persons with GMFCS level III might be overusing their walking resources in order to meet social expectations (e.g. the ability to cross the road on the green light, to walk together with non-disabled peers). There may also be other environmental factors, such as access to social services, home-based rehabilitation and physical training, in addition to body functions such as pain, fatigue and spasticity. Further, reduced balance and co-morbidities, such as epilepsy,

may also contribute to walking deterioration. Some of the shifts in GMFCS levels may be a result of deterioration, but can also be a matter of choice, as some might start using a wheelchair for energy conservation or pain relief over longer distances. In the younger age group (<25 years of age in 1999) 10% had experienced deterioration since 1999. This is a serious problem, as they have a long life ahead of them.

The most frequently reported characteristic of the deteriorated walking was reduced balance. Balance is an important part of walking function, and impaired balance has been found in children with CP (Brogren et al. 2001, Nashner et al. 1983, Burtner et al. 2007). Study I points towards a possible relationship between deteriorated walking and reduced balance, and further studies on balance and walking are needed.

The walking difficulties in spastic CP may be a continuing process over a longer period, from the first symptoms, which may be slight increased exertion, fatigue, pain, or increased stiffness during or shortly after walking, gradually leading to shorter and slower walks. At some point there may be a realisation that walking has deteriorated. Over time, walking distance, speed, and frequency may gradually decrease, until for some, walking is lost, and wheeled mobility becomes the only means of ambulation. This may lead to considerable activity restrictions and participation limitations, and may threaten the individuals' independence, and may have a negative impact on health due to a reduced physical activity level (FYSS 2008). It may therefore be important to identify walking deterioration at an early stage for preservation of walking.

People with CP have a motor disability that reduces their maximum potential (MAMP) and current movement capability (CMC) on many levels (Cott et al. 1995). The physiological reserves are also smaller than in the general population (Kemp and Mosqueda 2004). Therefore, even smaller changes in the MAMP and in the CMC may affect walking. As many different factors have an impact on walking and as walking is central within many different activities and important for independent living, as well as for the possibility to stay socially active, there is a need to focus on these factors in a life-span perspective, both in the clinical and the research communities.

6.1.2 Pain frequency, intensity and impact of pain

The number of pain sites and pain intensity on the VAS scale had not changed over the seven years, but pain frequency and impact of pain on daily life had increased. The relatively unchanged pain intensity level and number of pain sites agree with previous studies (Jensen et al. 2004, Malone and Vogtle 2009). The more often pain is experienced, the more it may interfere with daily activities. This may be the reason for the increased impact of pain on daily activities. Those who reported walking deterioration had higher pain scores on all scales than those without deterioration. This may be associated with a variety of different factors, such as spasticity, fatigue, muscle weakness, overuse, joint and muscle contractures, reduced cardiorespiratory fitness and physical endurance, osteoarthritis (OA) etc. Pain management is therefore an important element to consider when aiming at preserving walking in adults with CP.

A sensitization of the nociceptive afferent nerve fibres and the neurons in the dorsal horn that transfer pain signals from the muscles is a well known process that can take place in long-standing pain (Sessle et al. 1999). The sensitisation is often associated with hyperalgesia, which was demonstrated in individuals with chronic pain as enhanced responses to noxious stimuli (Giesecke et al. 2004). These mechanisms can also affect individuals with CP who have experienced pain over longer periods, and therefore contribute to chronic pain (Maslon et al. 2011). We have not examined the prevalence of either hyperalgesia or allodynia in this study.

6.1.3 Pain site prevalence

Back, neck, and foot/ankle pain were the most common pain sites in both unilateral and bilateral CP, which supports earlier findings (Schwartz et al. 1999, Murphy et al. 1995, Sandstrom et al. 2004). In bilateral CP, the reduced motor control often leads to excessive pelvic tilt and increased lumbar flexion/extension, lateral flexion and rotation during walking. This probably contributes to back pain, and the consequences may lead to lumbar spondylolysis (Murphy 2009, Harada et al. 1993). In unilateral CP, there is reason to believe that the asymmetry in motor control causes overuse of the non-affected side, asymmetrical joint loading and asymmetrical muscle activation patterns during walking, resulting in pain. Increased neck pain in spastic CP, may be due to compensatory increased cervical lordosis and spine movements, and was also reported by Ando and Ueda (2000). Back and neck pain are common in the GP, and a Norwegian population-based study found that 53% reported low back pain symptoms during the previous year (Natvig et al. 1995). The causal pathways for back and neck pain are probably different in the GP than in persons with CP. Musculoskeletal abnormalities may be a cause for the foot/ankle pain in CP. The prevalence of hip pain in bilateral CP was 43%, which is twice that in the GP (Natvig et al. 1995). The hip pain may be caused by the developmental changes common in bilateral CP, such as coxa valga, hip dysplasia and increased femoral anteversion. These skeletal changes in combination with altered muscle activity and joint mobility, increased muscle tone and abnormal joint loading may lead to secondary OA (Carter and Tse 2009, Murphy 2009). As foot and/or hip joints are considerably stressed during walking, pain in these joints may contribute to walking deterioration.

Females consistently reported more pain than males, which is in accordance with findings in the GP (Natvig et al. 1995, Wijnhoven et al. 2006). For example, females may be more willing to report pain; and may have greater exposure to risk factors than men (Wijnhoven et al. 2006). There are still many unanswered questions as to why these gender differences in pain perception and experience occur (Keogh 2006). These factors may also apply for the females with CP in this study.

Exertion was the most frequently reported cause of increased pain both in the back and in the neck, confirming the findings by Schwartz et al. (1999). This may indicate that overuse is a risk factor for pain, confirming the impact of physical components on pain. This may be in accordance with the hypothesis of Pimm (1992), who proposed overuse of resources as a cause of deterioration of functional skills. Physiotherapy was regarded as the most important pain-reducing factor in both back and neck pain. This confirms Schwartz et al. (1999), who found exercise and stretching to be pain-relieving factors in 92% and 89%, respectively.

Pain in adults with spastic CP may have different causes such as spasticity, contractures, abnormal joint loading, overuse of supporting muscles and ligaments due to reduced motor control and subsequent abnormal body segment alignment, reduced bone mineral density and inactivity (Vogtle 2009). It seems reasonable to believe that pain is an important factor in walking deterioration in an early age, with serious implications for activity and participation (Opheim et al. 2009, Jahnsen et al. 2004b). The need for improved pain management in this group is demonstrated by the findings of Studies I and II, and was also highlighted by Vogtle (2009) in a recent review article.

6.1.4 Number of pain sites and MCS/PCS

There was little, if any correlation between the number of pain sites and psychological health, as assessed with the MCS. This was in contrast to the general population (GP) in Norway where the number of pain sites was associated with both reduced physical and mental health (Kamaleri et al. 2008a). One reason for this finding might be that individuals with CP, having been disabled all their lives, are more “used to” having pain, or expect to have pain from time to time, thus having a different reference frame than the GP. Therefore, pain may not affect the psychological health in the same way as in the GP. Engel et al. (1999) found that pain-coping styles in individuals with CP were the use of task-persistence, diversion of attention and reinterpretation of pain sensations, which may be adequate coping styles for chronic pain (Jensen et al. 2011). Coping styles were not assessed in the present study. Drum et al. (2008) found that persons with disabilities, within the same self-rated health category as people without disabilities, had more days of poor physical and mental health. This suggested better tolerance of ill health among people with disabilities (Drum et al. 2008). This tolerance may, among other factors, be mediated by coping style. Another reason for the lack of correlation between MCS and number of pain sites may be that the expectations of pain improvement may be lower for people with a life-long disability who have had pain for many years. Pain in CP may mainly be of a physical origin, and be related to muscles and joints that are stressed or overused. These factors are well known to individuals with CP, which may make them easier to comprehend and incorporate in everyday pain compensation strategies. The negative correlation between the number of pain sites and PCS, although it was low, may confirm the physical components of pain in spastic CP.

6.1.5 Fatigue

Those reporting walking deterioration had higher physical fatigue scores than those not reporting. There were no changes in fatigue over the seven year period. This was rather surprising, but may be explained by the already high levels of physical fatigue (Jahnsen et al. 2003b) compared to the GP (Loge et al. 1998a), and also confirmed by Malone and Vogtle (2009). Those reporting walking deterioration did not have higher mental fatigue scores, but had significantly higher physical fatigue. This is suggesting that fatigue in CP is mainly of physical origin. Fatigue, as pain, may have multi factorial causes.

The experience of fatigue is individual, like pain, and cannot be measured directly. People with spastic CP have a motor disability that makes it more difficult to move and results in increased energy requirements during walking. Reduced muscle strength and cardiorespiratory fitness (Rimmer 2001) are common secondary

consequences and may contribute to physical fatigue. Coping with fatigue is also an individual matter, and is dependent on former experiences, adjustments, expectations and belief about capabilities (Bandura 1997). A person with a lifelong disability such as CP might expect to experience physical fatigue that is stronger than just tiredness from time to time. Because of these expectations, fatigue may not be experienced as overwhelming, and the impact on mental functions like concentration and memory may not become so great.

6.2 CASE-CONTROL STUDY

6.2.1 Gait analysis

We found no between-group differences in either work of walking or kinematic gait variables. The energy requirements during walking varied markedly across individuals, in both groups. Differences in work during walking may be experienced individually, and the assumed relationship between increased work of walking - fatigue - walking deterioration may also be mediated by factors such as cardiorespiratory fitness and muscle strength. Alternate walking patterns may also be energy-demanding (Gage et al. 2009). For example, crouch gait is energy-demanding due to high external hip and knee flexion moments, yet may not show increased W_{COM} . However, we did not find between-group differences in minimal knee flexion in stance. This suggests that other factors than work of walking may be important for self-reported walking deterioration.

The hypothesis that individuals reporting walking deterioration would have greater gait deviations shown by higher gait profile score (GPS) values was not supported. This has not been investigated previously. Self-reported walking deterioration may reflect different dimensions of walking other than kinematic gait quality. Reduced walking speed, distance and endurance, reduced confidence in walking over rough or slippery ground or stairs, or increased stiffness and pain during walking may all be perceived as deterioration, with associated activity limitations or participation restrictions. This may not be reflected in the highly standardised measures of short distance indoor linear walking as is the case in 3DGA and subsequent GPS values. The experience of walking deterioration may also be more dependent on changes in gait deviations, which must be elucidated in a longitudinal study.

The self-reported walking deterioration may also be a result of individual experiences and expectations in conjunction with the demands and strains on walking function. Others' expectations, different social and environmental factors, how the individual copes with the activity and participation restrictions caused by CP, and how the individual anticipates mobility will change over time may be important factors. Some may expect deterioration to occur, and not interpret changes as being worse than expected, and therefore report the situation as stable. Others, who anticipate stable walking, may interpret even small changes as deterioration. Changes in walking may also be experienced and interpreted differently due to differences in physiological reserve (Kemp and Mosqueda 2004), i.e. the difference between walking performance (CMC) and capacity (MAMP) (Cott et al. 1995). Those whose walking performance is closer to their maximal capacity may experience minor capacity reductions as deterioration, while those with larger reserves may not notice them.

Increased stiffness was a commonly self-reported cause for walking deterioration, typically related to the legs, back or upper body. There were no between-group

differences in overall muscle tone, supporting the view of multi-factorial causes for increased stiffness (Gracies 2005).

The Movement Analysis Profile (MAP) for the whole group had not been investigated previously, and indicated the largest deviations in the sagittal plane pelvis, hip and knee and foot progression angles. Increased pelvic tilt and reduced hip extension may be associated with hip flexion contractures, weak abdominal and/or hip extensor muscles and reduced SMC (Gage et al. 2009). Increased lumbar lordosis is common in spastic bilateral CP (Murphy 2009, Harada et al. 1993). Increased pelvic tilt may also be interpreted as a motor control strategy to increase pelvic stability by reducing the degrees of freedom (Woollacott et al. 1998). The knee deviations may be caused by reduced knee extension in mid-stance, as well as reduced and delayed maximal knee flexion in swing due to rectus femoris spasticity (Chambers et al. 1998). Foot progression angle deviations can be caused by both spastic and/or shortened triceps surae muscles causing both vaulting and rotation, bony torsions, or increased pes valgus with or without forefoot abduction. These foot deformities shorten the lever arm for propulsion at terminal stance and pre-swing (Gage et al. 2009), contributing to reduced ankle power at toe-off (Figure 8a-b).

The classification of gait patterns in adults with spastic bilateral CP (Rodda et al. 2004) has not been previously presented. In children with CP, the classifications are not always static and changes over time in both directions have been found (Rodda et al. 2004). Whether this classification is static or dynamic, or whether there is a natural deterioration towards Group IV in adults is unknown. Most were classified as either crouch or asymmetrical gait, and fifteen out of 32 and 8/32 limbs were crouch and apparent equinus, respectively. Crouch gait is an energy demanding gait, limiting walking range considerably (Gage et al. 2009), and apparent equinus puts considerable strain on the foot structures. The non-classifiable individuals had increased hip flexion and anterior pelvic tilt with relatively normal ankle and knee curves. These findings highlight the large gait deviations in this group. These deviations may indicate a need for individualised and goal-directed rehabilitation of walking.

6.2.2 Balance

Balance, and the association with walking in adults with CP, is a new research area which this study is the first to address. The main finding was that across all participants, there were considerable reductions in balance confidence, fear of falling and balance ability. However, there were no between-group differences in any of these variables. This may reflect a more complex interaction between walking deterioration and balance than originally hypothesised. In a Cochrane report, Howe et al. (Howe et al. 2007) concluded that, there was no consensus on the use of outcome measures of balance. This was one of the reasons for choosing to have a group discussion on balance problems, before choosing outcome measures for Study IV.

The results varied substantially in both groups, which may reflect the heterogeneity of spastic CP. If there are changes in balance ability over time, these may be experienced differently across individuals, different social roles and exposure to different contexts, as well as personal factors put different demands on both balance and walking. A small change in balance might therefore have a considerable impact on walking for some with high functional demands and expectations, whereas for

those with smaller demands these changes might go unnoticed. Possible changes in balance may be more important for the experience of walking deterioration, than the balance at a given time.

Across the whole group, BESTest showed reductions in all subsystems of balance, and the postural responses and anticipatory adjustments seemed to be most affected. The total BESTest were considerably lower than in Lord et al. (2010) in healthy elderly. Anticipatory adjustments and postural responses are a result of life-long motor learning and experience, and motor learning is a result of frequency, intensity and variation of both the complexity of the movements and the contexts in which they take place (Shumway-Cook and Woollacott 2007). The reduced postural responses are in accordance with Gage (2009) (p107-128) and Burtner et al. (2007). The ability to take a rapid step to regain balance is a crucial postural response to avoid falls, and Lajoie and Gallagher (2004) found significantly longer reaction times in elderly fallers. The high number of falls in this group may partially be caused by the reduced postural responses.

Anticipatory postural adjustments reflect the ability to anticipate the destabilising effect of movements (Horak et al. 2009), and the reductions were probably caused by motor learning and reduced motor control. In addition, people with spastic CP are often less active in play and sports than other children and young adults (Michelsen et al. 2009, van der Dussen et al. 2001). Because of these two factors, people with spastic bilateral CP may be said to have a double disadvantage. Increased muscle tone, antagonistic co-activation (Burtner et al. 1998), maybe as a strategy to cope with reduced postural control (Carlberg and Hadders-Algra 2005), a top-down muscle activation (Nashner et al. 1983), a reduced ability to modulate EMG amplitude (Brogren et al. 2001) and muscle weakness and joint impairments are likely to be contributing factors.

The overall balance confidence was lower than in community-dwelling elderly (Wrisley and Kumar 2010), and at the same level as in elderly after hip fractures (Sihvonen et al. 2009). Fear of falling showed a similar trend as balance confidence and could be classified as moderate (Delbaere et al. 2010), slightly lower than in a group of people with spinal cord-injuries (Wirz et al. 2009), and at about the same level as elderly treated for fall-related fractures (Nordell et al. 2009). The high number of falls and the risk of an injury, reduced balance confidence and fear of falling may substantially restrict both social and community participation for these relatively young people. Fatigue may also have an impact on both balance and falls, but this relationship was not assessed in this study.

The relationship between balance confidence and balance ability, showed a moderate positive correlation for the postural responses, sensory orientation, stability in gait and total BESTest score. Because of the motor control problems being more distal (Nashner et al. 1983, Gage et al. 2009), there may be a reduced ability to use an ankle strategy to regain balance. This, in addition to the reduced postural responses, creates a dual problem, as both the most adequate balance adjustment strategy and the ability to take rapid steps to regain balance are reduced. Low balance confidence when using escalators and walking on slippery surfaces may confirm this. The positive correlation between balance confidence and several tests of balance ability suggests that the participants had a realistic perception of their own balance.

6.3 GENERAL ASPECTS

The underlying intention of this thesis has been to focus on the walking problems that a considerable number of people with spastic CP experience. Therefore, gaining more knowledge about the critical variables and the relationship between them may assist both professionals and patients in choosing treatment and rehabilitation interventions that target this specific problem. People with spastic CP have a lifelong disability that leads to activity limitations and participation restrictions of various kinds, and additional deteriorations may impede independence. The relationship between impairments of body functions and structures, activity and participation restrictions, and the disability are complex (Østensjø et al. 2004, Holsbeeke et al. 2009).

Ageing with CP has been addressed by Kemp and Mosqueda (2004), and one central concept is the physiological reserve. This relates to the ability to increase organ function, or sustain stress and strain, if demanded. In people with CP, the upper limit of the physiological reserve is often lower than in the GP. With increasing age, the physiological reserve is reduced for all, but as disabled individuals have a lower upper limit, the reserves are smaller, and the difference between daily functioning and the upper limit becomes smaller. For people with a chronic neurological disorder, with already marginal, but important walking function and small reserves which may have already decreased since adolescence, additional problems may lead to increased walking deterioration and subsequent activity and participation restrictions (Kemp and Mosqueda 2004). Reductions in reserves may result in increased physical fatigue. Increased energy expenditure during walking was found in children with CP (Dallmeijer and Brehm 2011, Perry 1992, Duffy et al. 1996). There is reason to assume the same in adults as well. Mattsson et al. (1990) found oxygen cost during level walking in adults with spastic paraparesis to be double that of healthy subjects (Linnarsson et al. 1989). The energy expenditure during walking in children with CP was close to the recommended values for cardiorespiratory fitness training (Dallmeijer and Brehm 2011, Åstrand et al. 2003), and thus might be a factor in early onset of fatigue. Van der Dussen et al. (2001) found reduced integration in sports activities, and Jahnsen et al. (2003a) found that almost half were regularly physically active, but 39% were inactive. The cardiorespiratory fitness level in adults with CP varied between 23-61% of non-impaired subjects (Rimmer 2001), confirming reduced fitness. Therefore, a prevention of additional declines of physical activity and fitness levels may be important, as reduced physical activity have several negative health consequences such as osteoporosis and increased risk for cardiovascular diseases (FYSS 2008).

The above findings, together with the findings of the present studies, point towards reduced cardiorespiratory fitness and physical activity levels, reduced muscle strength and increased pain and fatigue among those with deteriorated walking. This, in addition to the considerable gait deviations, especially the crouch gait seen in almost half of the limbs, and the reduced balance confidence, balance ability and the high number of falls may be central problems that lead to activity and participation restrictions for these young individuals.

It is important to remember that the CP does not disappear, and the disability will be there for life. However, coping with the consequences and complications may become better and adapted to the whole life situation. People with CP also develop other diseases. Some of these symptoms may be overlooked as being expected sequelae of CP (Murphy 2009), or be hidden by the more eminent abnormal

movements and communication difficulties. Blaming all ailments, pain and functional reductions on the CP and its consequences and complications may cause health workers to overlook underlying conditions of neurological or other, and malignant origin. It is therefore important to exclude other possible medical causes of increased muscle tone, pain and fatigue and reduced walking and balance.

6.3.1 Movement Continuum Theory and ICF

Referring to the MCT (Cott et al. 1995), the measurements in these studies have taken place on several different levels, such as tissue (e.g. bone deformities), organ system (e.g. muscle strength), body part (e.g. kinematics of the lower limb), body (e.g. gait pattern), the individual in the environment (e.g. BP from SF-36 or FMS), and the individual in society (e.g. PCS and MCS). The balance ability and FMS may inform about current movement capability (CMC). The maximum achievable movement potential (MAMP) was assessed in muscle strength and joint ROM, and some of the balance ability tests may also assess MAMP. To formally assess the MAMP at other levels or to assess the interaction between levels were beyond the scope of this thesis.

MCT and ICF have several similarities (Cott et al 1995, WHO 2001). For example both have levels of body functions and the individual in society. Both also consider the possible discrepancy between the MAMP and the CMC in the individual. There is also a great emphasis on the interaction between the different levels, and an understanding of the non-hierarchical and a bio-psycho-social model of disability (WHO 2001). One main difference between them is that ICF does not classify the preferred movement capacity (PMC). The capacity modifier in ICF may not be equal to the MAMP from MCT, and therefore not reflect the potential. These distinctions may be important, because the MAMP reflects what you can do now, if you want in your own environment and with the aids you normally use. How fast you can walk or run, or how long you can stand on one leg, etc. This is in contrast to the capacity modifier of activity in ICF which is supposed to be assessed “naked” in a standardised environment.

As most people do not run as fast as they can all the time, the CMC reflects the level of daily functioning, and the PMC the level that the individual wants to function at (Cott et al. 1995). If a discrepancy between these two levels arises, there are two ways to minimise the difference, either by lowering the PMC or by raising the CMC (or both). In adults with spastic CP who are reporting walking deterioration, there may be a PMC/CMC discrepancy, and the PMC may be higher than the CMC. The PMC in walking may reflect the function that the individual has had over many years. Increased strain of walking (Dallmeijer and Brehm 2011), reduced physical activity level (Nieuwenhuijsen et al. 2011), reduced cardiorespiratory fitness (Rimmer 2001), muscle strength, increased muscle tone, increased body weight, musculoskeletal pain and fatigue, reduced balance and fear of falling, may all be contributing factors to a reduced CMC. PMC, on the other hand is not only a physical construct, but may also be thought of as incorporating mental and/or psychological factors (Cott et al. 1995), thus, a result of experiences and past levels of functioning, as well as the influence from the physical and social environment. When an individual experiences walking deterioration (reduced CMC) the PMC may remain at a higher level for a long time for some people, because it may be difficult to accept a permanent deterioration. Others may be less distressed by this, and much quicker adapt the PMC to a new level, or commence the necessary actions to lift their CMC level. Not all persons

reporting walking deterioration will have a PMC/CMC discrepancy. In these persons an adaptation of PMC/CMC levels may have taken place. A constant PMC/CMC discrepancy might be felt as frustrating and negative, and an adaptation to a new PMC/CMC level may be regarded as successful coping.

Within the ICF domains (WHO 2001), walking may be regarded as either a body function and/or activity. Gait patterns are classified as body functions, and 3DGA thus reveals impairments. Walking tests such as the 6 min walk test reflects an activity and is often used as a test of capacity. The modifier of capacity is different from the modifier of performance, which reflects the activity limitations within the individual's own natural environment. The capacity reflects the activity limitations when being "naked" and in a standardised environment, thus removing the potential positive effect of technical aids, or a potential negative effect of the environment, but environmental modifications may also increase capacity (WHO 2001). The concept of capability was used by Holsbeeke et al. (2009) to describe what the individual is able to do in his own environment if he/she has to. This adds a level between the performance (what he/she usually does), and the capacity (maximum level), and may be a useful addition in understanding the interplay between impairment, disability and function.

6.4 METHODOLOGICAL CONSIDERATIONS

The generalisability of these studies must be discussed in three stages. First, the representativeness of the participants eligible for the follow-up study, i.e. the representativeness of the participants with spastic unilateral and bilateral CP without intellectual disabilities in Jahnsen's (2004) study with respect to the whole population of spastic CP in Norway. The original study succeeded in recruiting a good representation of the whole population (Jahnsen 2004), and the non-responder analysis showed no differences with regard to diagnosis, age and geographical representation. Distribution of sub-diagnoses also corresponded well with international epidemiological studies of CP. There is no central register for CP in Norway, and those who had not been in contact with the habilitation services were not reached (Jahnsen 2004). The follow-up study also reflects this. The second stage concerns the representativeness of the participants in the follow-up study in 2006, with respect to those eligible for the study. Only persons with spastic CP were included in the follow-up. A non-responder analysis was carried out, and showed no differences in age, GMFCS level or gender between responders and non-responders. Therefore, the participants in the Studies I-II may be representative of adults with spastic CP, without intellectual disabilities, with mild to moderate severity of CP. The third stage concerns the representativeness of the participants in Studies III and IV. They were recruited from the follow-up study based on specific criteria, and thus a selected sample, and not a self-selected or a convenience sample. One of the strengths was the recruitment of non-hospitalized individuals, and recruiting community-dwelling individuals from all over the country as in this study, may increase representativeness. More than half of the group were working, and had more than 12 years of education. Therefore, they may be broadly representative for adults with spastic bilateral CP, GMFCS level II.

None of the methods in Studies III-IV, i.e. 3DGA, balance confidence and balance ability had been formally tested for reliability and validity in adults with spastic bilateral CP before. To address this shortcoming, we conducted an intra-rater

reliability study on 12 adults with spastic bilateral CP. The reliability of the balance confidence and the balance ability tests were comparable to studies of individuals with stroke and a mixed neurology group (Botner et al. 2005, Horak et al. 2009). The error of measurement of the 3DGA kinematics was within the limits suggested in the systematic review by McGinley et al. (2009) of 5°.

The 3DGA was performed in a laboratory and assessed impairments of body functions. However, walking as an activity involves walking in different contexts and for different purposes, and performance in these settings is considerably different from that in the laboratory. Thus, only some aspects of walking are captured with 3DGA, and walking is substantially more than gait in a laboratory. On the other hand, walking a short distance on an even surface indoors, without any visual or audio distractions may be argued to be considerably easier than walking outdoors. Thus, the gait impairments revealed by 3DGA, may be even greater in other, and more demanding settings. Walking was also described with the FMS, taking into account both the walking distance and the degree of support needed, thus indicating performance in daily life.

The ABC, FES-I, BESTest and FSST were chosen based on the balance problems highlighted in the group discussion and from clinical experience. The ABC (Powell and Myers 1995) and FES-I (Yardley et al. 2005) are considered to reflect both the balance confidence and the fear of falling during different daily activities. Both of these scales were included, because although they have similarities, the questions are radically different. ABC asks about confidence of not falling, which may be said to represent the belief about capabilities, thus, the self-efficacy (Bandura 1997), and the positive coping resources related to balance. FES-I asks about the fear of falling, and thus the negative side, or maybe the absence of balance self-efficacy. Therefore these two scales may be regarded as reflecting two different sides of the same coin (balance), both the positive side (confidence in keeping the balance), and the negative side (fear of not keeping the balance). These scales were negatively correlated to each other, thus supporting this view. ABC correlated with balance ability, suggesting an internal consistency, and indicating concurrent validity, also found by Horak et al. (2009).

The measurements of balance includes the complex processes that are involved in integrating both neural and musculoskeletal body functions and structures, as well as the involved task and the context in which this task takes place, thus requiring a comprehensive approach (Mancini and Horak 2010, Matsumura and Ambrose 2006). Direct measurement of body sway can be made from centre of pressure (COP) readings on a force plate (Piirtola and Era 2006). This may be of limited relevance for how the balance affects the performance of functional tasks such as standing up, turning, walking etc (Hughes et al. 1996). Many clinical tests of balance are therefore indirect measurements of balance, measuring a functional task challenging the balance, with the assumption that reduced balance leads to reduced task performance (Howe et al. 2007). There was also no consensus about which tests of balance ability are the most effective, as concluded in the Cochrane review by Howe et al. (2007). Another shortcoming of the common clinical tests of balance, is the inability to differentiate between the different body functions and structures that are involved in balance (Mancini and Horak 2010, Horak et al. 2009). It may therefore be difficult to interpret the results, and define targeted therapeutic exercises for the individual (Horak et al. 2009). Thus, Horak et al. (2009) have, partly on the basis of existing clinical tests, and partly of newly developed tests, designed the BESTest. This test

was designed to evaluate six different subsystems of balance, and reveal the subsystem(s) that is affecting balance most. The BESTest cover several of the balance problems highlighted in the group discussion.

Spasticity in the lower limbs was present among all individuals, but with individual differences, and probably had an impact on all subscales of BESTest. The overall score on the BESTest may to some degree reflect the overall motor impairments in the individual, which may also be affected by spasticity. However, in these studies, it was the consequences of spasticity as one of several contributing factor to reduced balance and impaired gait, and possibly to walking deterioration, and not the spasticity *per se*, which was the object of study.

Studies I and II have a fairly large number of participants (N=149), and the conclusions may be drawn with relative certainty. In studies III and IV, only 16 participants fulfilled the inclusion and exclusion criteria, which was less than had been hoped. The inclusion and exclusion criteria may have been too narrow, but were chosen because of the known heterogeneity in the CP group, and because the targeted group were assumed to reflect those at risk of walking deterioration. Because of the low number of participants in studies III and IV, a conservative statistical approach was chosen, and non-parametric statistics was used.

6.5 STUDY LIMITATIONS

Self-report questionnaires may be biased by factors such as recall, interpretation, motivation and emotion. Expectations regarding research outcome or regarding what is expected may also affect the results. However, in a large group, these differences tend to level out as they affect the groups randomly, and thus do not cause systematic errors. Health researchers have found agreement between self-rated health and measured health and mortality (Burström and Fredlund 2001, Larue et al. 1979), thus indicating that self-report may be used to obtain valid and reliable data.

The main limitations of Studies III and IV were the small sample size and the cross-sectional case-control design. If there were changes in balance and gait variables over time, these may be more important for walking deterioration than gait and balance at a given time. The group may be considered to be broadly representative of ambulant community-living adults with mild to moderate spastic bilateral CP, but the results need to be confirmed in other studies, and the results cannot be generalised to a larger group.

7 CONCLUSIONS

- Increased prevalence of walking deterioration was reported, mostly in spastic bilateral CP in the 35-40 years age group, and in higher GMFCS levels
- Increased pain frequency and impact of pain on daily activities were reported, but unchanged physical and mental fatigue over a seven-year period.
- Walking deterioration was associated with higher levels of pain and physical fatigue.
- Back, neck and foot/ankle were the most common pain sites, and increased prevalence of back and neck pain over a seven-year period was found. Pain was more common in bilateral CP and in females.
- Those reporting more pain sites did not show worse psychological health, which is in contrast to the general population, and suggesting adequate coping strategies.
- Walking deterioration could not be explained by differences in work of walking or kinematic gait variables.
- Gait was characterised by increased pelvic tilt, hip and knee flexion and deviating foot progression angle, and as almost half of the limbs were classified as crouch pattern, this was suggestive of increasing walking difficulties.
- Walking deterioration could not be explained by differences in balance confidence, fear of falling or balance ability.
- Balance confidence and fear of falling were at the same level as in elderly after hip fractures. The high number of falls and the reduced balance ability especially during activities requiring postural responses and anticipatory adjustments, may lead to activity and participation restrictions for these young individuals.

8 CLINICAL IMPLICATIONS AND FUTURE STUDIES

- These studies demonstrate a need to establish evidence-based rehabilitation in a life-span perspective for this group. Intervention studies directed at managing and treating increased pain and fatigue are needed. Evidence-based physiotherapy interventions for improving or preserving walking and balance are also needed.
- Before full evidence has been established, structured, evidence-based/best practice follow-up programmes in specialised rehabilitation settings should be implemented. Individualised, goal-directed interventions in a life-span perspective need to be developed, aiming at preventing secondary complications and activity/participation restrictions.
- Walking in adults with spastic CP should be studied along three main lines, preferably longitudinally:
 - The natural history of kinematic and kinetic gait variables, as well as of balance in order to understand the causes and mechanisms underlying walking deterioration
 - The effect of rehabilitation interventions on kinematic and kinetic gait variables, as well as on walking and balance in relation to activities and participation.
 - Qualitative studies exploring the meaning and interpretation of walking deterioration
- The utility of the classification of sagittal gait patterns with defined biomechanical characteristics, postulated sequelae and management suggestions for children needs to be investigated for adults with spastic CP.

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