

Linköping Studies in Health Sciences, Thesis No. 91

# **Adults with Cerebral Palsy**

## **- living with a lifelong disability**

**Karin Sandström**



**Linköping University**  
**FACULTY OF HEALTH SCIENCES**

Division of Physiotherapy  
Department of Medical and Health Sciences  
Linköping 2009

©Karin Sandström, 2008

[Karin.Sandstrom@liu.se](mailto:Karin.Sandstrom@liu.se)

<http://www.imh.liu.se>

Electronic publication of the thesis:

<http://urn.kb.se/resolve?urn=urn:nbn:se:liu:diva-15771>

Cover illustration: Benita Rennes

Published article has been reprinted with the permission of the copyright holder.

Printed in Sweden by LiU-Tryck, Linköping, Sweden, 2008

ISBN 978-91-7393-731-3

ISSN 1100-6013

# CONTENTS

<b>ABSTRACT .....</b>	<b>1</b>
<b>LIST OF PAPERS.....</b>	<b>3</b>
<b>ABBREVIATIONS .....</b>	<b>4</b>
<b>INTRODUCTION.....</b>	<b>5</b>
CEREBRAL PALSY - DEFINITION AND CLASSIFICATION .....	5
PREVALENCE AND LIFE EXPECTANCY.....	7
LIVING WITH CP AS AN ADULT.....	7
PHYSICAL ACTIVITY AND PHYSIOTHERAPY .....	9
RATIONALE FOR THE THESIS .....	10
<b>AIMS .....</b>	<b>11</b>
SPECIFIC AIMS.....	11
<b>METHODS .....</b>	<b>12</b>
SUBJECTS .....	12
PROCEDURE .....	14
<i>Sample I (paper I).....</i>	<i>14</i>
<i>Sample II (paper II and III).....</i>	<i>15</i>
STATISTICS AND ANALYSES .....	16
<i>Paper I.....</i>	<i>16</i>
<i>Paper II.....</i>	<i>16</i>
<i>Paper III.....</i>	<i>17</i>
VALIDITY (PAPER II AND III).....	17
ETHICAL CONSIDERATIONS .....	18
<b>RESULTS.....</b>	<b>19</b>
GROSS MOTOR FUNCTION CLASSIFICATION SYSTEM (GMFCS) AND RELATIONS TO INSTRUMENTS MEASURING HEALTH AND FUNCTIONING (PAPER I).....	19
DESCRIPTIONS OF FUNCTIONING AND HEALTH (PAPER I).....	19
<i>Changes in gross motor function from adolescence to adulthood .....</i>	<i>19</i>
<i>Activities of daily living and locomotion.....</i>	<i>20</i>
<i>Passive range of motion (PROM) and pain.....</i>	<i>21</i>
<i>Health related quality of life.....</i>	<i>22</i>
THE LIVED BODY- EXPERIENCES OF LIVING WITH A DISABILITY (PAPER II).....	23
<i>Perceptions.....</i>	<i>23</i>
<i>Strategies .....</i>	<i>25</i>
PREREQUISITES FOR CARRYING OUT PHYSIOTHERAPY AND PHYSICAL ACTIVITY (PAPER III) .	26
<i>Being enjoyable .....</i>	<i>27</i>
<i>Giving effects.....</i>	<i>27</i>
<i>Being comprehensible.....</i>	<i>27</i>
<i>Being integrated in daily life .....</i>	<i>27</i>

<i>Supportive health care with competent professionals</i> .....	27
<b>DISCUSSION</b> .....	<b>29</b>
METHODOLOGICAL CONSIDERATIONS .....	29
DISCUSSION OF RESULTS .....	31
<i>Deterioration in gross motor function according to GMFCS</i> .....	32
<i>The 'dys-appearing' body and its impact on daily activities and self-perceived health</i> ..	32
<i>Self-image</i> .....	35
<i>Being autonomous</i> .....	37
<i>Physical activity and physiotherapy</i> .....	38
<i>Transitions in life and strategies to meet them - implications for professionals</i> .....	41
<b>CONCLUSIONS</b> .....	<b>44</b>
<b>SAMMANFATTNING</b> .....	<b>45</b>
<b>ACKNOWLEDGEMENTS</b> .....	<b>47</b>
<b>REFERENCES</b> .....	<b>49</b>

## **ABSTRACT**

Although Cerebral Palsy (CP) is a lifelong disability, the research has mainly focused in children with CP. However, in recent years new studies have examined the different aspects of being an adult with a congenital disability.

The overall aim of this thesis is to contribute to increased knowledge and understanding in living with cerebral palsy as an adult, with special focus on the lived body and physiotherapy/physical activity.

The thesis is based on two populations. The first population (study I) consists of 48 individuals with a variation in gross motor function equivalent to the general CP-population, including all five levels according to a gross motor classification (Gross Motor Function Classification System, GMFCS). The second population (study II and III), consists of 22 individuals, representing level II to IV according to the gross motor classification (GMFCS).

Study I comprises structured questions and clinical investigations with well-recognized assessment instruments and are analysed according to quantitative methods. Study II and III are based on in-depth interviews and are analysed using qualitative methods.

The overall result is that there is a deterioration of motor function in many individuals already early in adulthood. These deteriorations are preceded and/or followed by musculoskeletal problems such as pain and limited range of motion, often in combination with fatigue. Despite deteriorations many people maintain daily personal activities, but are often forced to prioritize among societal activities.

The deterioration is often perceived as slow and imperceptible, but it also results in a change in self-image with new thoughts about otherness and being different. This can be experienced in relation to activity limitations with difficulties in taking part in other people's activities, but also in relation to attitudes and treatment from other people. The process of deterioration also influences autonomy, with limited ability to determine one's own daily life.

Important prerequisites for carrying out physical activity in a longer perspective is that it has to be enjoyable, give effects, be comprehensible and

integrated in daily life. In addition it is important with support from competent professionals in health care. Experiences of these prerequisites vary and, above all, in adulthood the lack of competent support is a factor which is illuminated of the majority of the interviewed and assessed individuals.

## LIST OF PAPERS

- I. Sandström K, Alinder B, Öberg B. Descriptions of functioning and health and relations to a gross motor classification in adults with cerebral palsy. *Disability and Rehabilitation*, 2004; 26 (17): 1023-1031
- II. Sandström K. The lived body – experiences from adults with cerebral palsy. *Clinical Rehabilitation*, 2007; 21 (5): 432-441
- III. Sandström K, Samuelsson K, Öberg B. Prerequisites for carrying out physiotherapy and physical activity – experiences from adults with cerebral palsy. *Accepted for publication in Disability and Rehabilitation*.

## **ABBREVIATIONS**

ADL	Activities of Daily Living
CP	Cerebral Palsy
EQ-5D	European Quality of Life 5 Dimensions Questionnaire
FIM	Functional Independence Measure
GMFCS	Gross Motor Function Classification System
IADL	Instrumental Activities of Daily Living
IAM	Instrumental Activity Measure
PROM	Passive Range of Motion
S-COVS	Swedish – Clinical Outcome Variable Scale
VAS	Visual Analogue Scale



## INTRODUCTION

This thesis deals with different aspects of living with cerebral palsy, with focus on adulthood. The starting point was based on my clinical experiences as a physiotherapist, including histories conveyed in the meetings with adults living with cerebral palsy. At the same time there was an ongoing debate about training and follow-up for adults with cerebral palsy. The County Council of Östergötland wanted to gain more knowledge about these aspects addressing the role within the health care system. This led to an investigation that resulted in descriptions about functioning and health, as well as new research questions that addressed how people live with cerebral palsy, from a life perspective.

### **Cerebral Palsy - definition and classification**

Cerebral palsy (CP), a group of neurological conditions, is a non-progressive disturbance in the developing nervous system.<sup>1,2</sup> Although brain damage is a one-time event, impairments often change over time.<sup>1</sup> CP is defined according to movement and posture, but is often accompanied by other disturbances of sensation, cognition, communication, perception, behaviour and/or seizure disorders.<sup>2</sup> The Surveillance of Cerebral Palsy in Europe (SCPE), a network with registers from 14 centres in eight countries, reported that 31% of children with CP had intellectual impairments, 11% visual impairments and 21% epilepsy.<sup>3</sup> In a heterogeneous cohort of children with CP, a Swedish study showed that 40% had learning disability, 35% epilepsy and 20% visual impairment.<sup>4</sup> The condition is very heterogeneous and the effects of CP vary widely from person to person.

There are two ways to classify CP according to motor disorders. The most common classification divides CP into subgroups with predominant motor disorder (spastic, ataxic or dyskinetic) and topographical distribution (hemiplegia, diplegia or tetraplegia).<sup>5</sup> Recently, the European network (SCPE) agreed to use the concepts unilateral and bilateral according to body location.<sup>3</sup> The most frequent subtype according to SCPE is bilateral spastic CP (55%), followed by unilateral spastic CP (29%), dyskinetic CP (7%) and ataxic CP (4%).<sup>3</sup>

Another system to classify motor function, developed by a Canadian research group<sup>6</sup> is the Gross Motor Function Classification System (GMFCS), a system that is based on self-initiated movements, with emphasis on walking and sitting. GMFCS, a five-level classification (Table 1) classifies the usual performance in daily life. Distinctions between the different levels are based on functional limitations and the need for assistive technology, including mobility devices and wheeled mobility.

The classification consists of four age-bands: between 0-2; between 2 and 4, between 4 and 6; and between 6 and 12. The 6-12 version has been used in the adult population and considered a reliable classification instrument even for adults with CP,<sup>7</sup> although this was not the case when the first study in this thesis was performed. Recently, a new age-level, 13-18 years, has been developed,<sup>8</sup> but it is not used in this thesis.

*Table 1. GMFCS levels Shortened version, 6- 12 years (Palisano et al. 1997).*

---

<b>GMFCS levels</b>	<b>Gross Motor Function</b>
<b>I</b>	Walks without restrictions; limitations in more advanced motor skills
<b>II</b>	Walks without assistive devices; limitations in walking outdoors and in the community
<b>III</b>	Walks with assistive mobility devices; limitations walking outdoors and in the community
<b>IV</b>	Self-mobility with limitations; children are transported or use power mobility outdoors and in the community
<b>V</b>	Self-mobility is severely limited with the use of assistive technology

---

## Prevalence and life expectancy

In Sweden, CP is the most common cause of motor disability in childhood with a prevalence of 1.9-2.7/1000 births.<sup>9-11</sup> In a cohort in the UK, 85% of adults with CP were still living at age 50; in the general population 96% of all people survive until the age of 50.<sup>12</sup> Severe motor impairment increases the risk of early mortality<sup>13, 14</sup> and more than a third of people with a severe disability die before 30 years of age.<sup>15</sup> However, Strauss et al.<sup>16</sup> have showed that during a 20-year period (1983-2002) the survival in people with severe CP has increased by five years. This increase of survivability may be due to better nutrition and ventilation techniques. Consequently, the majority of people with CP reach adulthood<sup>15</sup> and the relative risk of death compared with general population has decreased with age; however, after 50 years of age there is some indication that the survivability of people with CP decreases relative to the general population.<sup>12</sup> Survival curves for 904 persons with CP between age 60 and 75 show that mobility function is a predictor for life expectancy in older people.<sup>17</sup>

## Living with CP as an adult

Because CP is a congenital disease, research has focused on childhood issues. However, in recent years there has been more attention paid to living with CP in a life perspective and especially the transition from adolescence into adulthood.<sup>18,19</sup> Environmental barriers such as poor transition to adult health services, poor accessibility, and lack of support are stressed, as well as problems in connection with people's ability to communicate their own needs and ability to make their own decisions.<sup>18-23</sup>

In the 1990s, studies began to explore how adults deal with CP. These studies focused on health issues such as medical and functional status.<sup>24-28</sup> They reported increasing musculoskeletal problems in adulthood and described medical problems with seizures, nutrition, aspiration, hearing and visual impairments. Today research has noted that in adulthood secondary problems – such as pain, fatigue, joint deformities and other musculoskeletal problems – often increase.<sup>25, 29-33</sup> These problems often lead to activity limitations, because of reduced mobility.<sup>34-37</sup> Many of these conditions are the result of straining oneself in order to manage daily life and problems finding a balance between overuse and inactivity.<sup>35,37</sup> People with CP generally notice these effects of

ageing earlier than non-disabled people. Increasing problems with reduced mobility leads to problems with daily activities, especially in instrumental activities of daily living (IADL);<sup>38</sup> however, despite mobility limitations, many adults with CP have a high level of independence in performing activities.<sup>39</sup>

Recently, studies also have focused on participation and social issues. The results differ, but the general message is that social participation, sexual relationships, employment and leisure activities are restricted among many youths and young adults.<sup>40-44</sup> Severity of physical and/or cognitive impairment are predictors for limited participation,<sup>41,45,46</sup> but limited participation is not necessarily synonymous with a poorer quality of life.<sup>47</sup>

As described above, there are documented problems in health status in adults with CP and they increase with ageing. According to self-perceived health there are more contradictory results, however indicating that functional level is not a predictor for low self-perceived health or quality of life.<sup>27,48,49</sup> Few studies examine self-perceived health or quality of life among adults with CP. One study, however, that focused mainly on persons with developmental disabilities (including adults with CP) showed that the greatest correlate to quality of life was accessibility to transportation and meaningful employment.<sup>50</sup> Some studies also show that pain, falling stamina and deterioration of function have a negative impact on quality of life.<sup>48, 51</sup>

Issues about health and living with a disability can also be explored using phenomenological theories, whereof Husserl is the most important predecessor.<sup>52</sup> Phenomenology provides us with a language that is based on lived experiences and formulated by Husserl as 'back to the things themselves'. Those lived experiences are expressed as feelings, thoughts and actions<sup>53</sup> and elucidate the meaning of everyday life. The individual's experiences and interpretation of his or her situation can give deeper understanding of everyday life and living with a lifelong disability.

Experiencing everyday life can also be expressed by the concept 'the lived body', as described by the French philosopher Merleau-Ponty.<sup>54</sup> It is through our body we have access to the world; the body 'understands', and 'inhabits' the world. This means that the lived body represents my unique 'being-in-the-world', but you can also observe and talk about the body.<sup>54</sup> In this way, the body can both be a subject (I am my body) and an object (I have a body), a relation between subjective experience and reflexion.

Literature about adults living with CP is primarily descriptive in nature and lived experiences are still sparsely described. Only two studies with this 'insight perspective' were found focusing on adults with CP. A study about experience of living with constant pain showed how pain consumed the person's thoughts, time and energy.<sup>55</sup> Another study containing interviews with older adolescents illustrated important factors that influenced self-image, such as being believed in, believing in yourself and being accepted by others.<sup>56</sup> To gain more knowledge about living with a lifelong disability, more research should focus on the individuals' own views.

## **Physical activity and physiotherapy**

There are many interpretations of physical activity, from doing daily activities to specific training schedules. A broad definition is 'any bodily movement produced by the contraction of skeletal muscles that increases energy expenditure above basal level'.<sup>57</sup> Physiotherapy that addresses both children and adults with CP often has a broad focus. It contains a spectrum of physical activity that includes individual training with a physiotherapist, physical training in different settings and varying physical activities in daily life. These boundaries are hard to establish, which also agree with the definition of physiotherapy identified by the World Confederation of Physical Therapy (WCPT) as 'identifying and maximizing movement potential, within the spheres of promotion, prevention, treatment and rehabilitation'.<sup>58</sup> This thesis alternates between the concepts physical activity and physiotherapy, which includes daily activities, leisure activities, exercise programs and more specific training programs.

Today, it is generally accepted that physical activity promotes health. To this end, there is a large amount of good information about performing physical activity. Advices how to perform different kind of activities are presented in different guidelines, even adapted to persons with disabilities;<sup>59</sup> however, it is difficult to find such guidelines for people with CP and the research literature is limited according to effects of physical activity in adults with CP. A few papers report benefits from participating in strength training,<sup>60-63</sup> and a pilot study with seven adults with CP showed improvements in cardio respiratory fitness after an 8-week program with a bicycle ergometer.<sup>64</sup> The great majority of the literature that has focused on children with CP shows effectiveness of strength-training<sup>65</sup> and intensive training programs.<sup>66</sup> Even if it is an

assumption that adults with CP will benefit from physical activity, there is a need for more investigations about effects and performance.<sup>67, 68</sup>

Consumption of physiotherapy and other physical activities are reduced in adulthood compared to childhood.<sup>69,70</sup> Environmental barriers such as accessibility, cost, and lack of physical assistance<sup>67,71,72</sup> as well as lack of initiative and motivation are reported.<sup>69</sup> With the exception of one study, which showed that ten persons with CP who participated in strength training in a group experienced this as enjoyable,<sup>63</sup> no studies about experiences of physical activity/physiotherapy in adulthood was found. More knowledge about one's own experiences and thoughts about being physically active in a longer perspective are needed in order to support and find motivation for active lifestyles.

## **Rationale for the thesis**

Clearly, there are many aspects related to adulthood and ageing with CP. So far, studies have mainly focused on deterioration in mobility and secondary musculoskeletal problems. Participation and social issues are more sparsely described and only a few studies deal with psychological dimensions such as mood, self-image or self-esteem. No studies about life experiences living with cerebral palsy and being a physical active person were found. It seems that there is a need for more knowledge about living with CP from a life perspective. This thesis deals with living with a lifelong disability with a major focus on the person's own experiences to provide more knowledge and understanding about how to enhance support of adults living with CP in varying life situations.

## AIMS

The overall aim of this thesis is to contribute to increased knowledge and understanding of living with cerebral palsy as an adult.

### Specific aims

To investigate how Gross Motor Function Classification System (GMFCS) relates to other established measures of functioning and health in an adult population with CP (paper I)

To give a general description of adults with CP according to

- functioning and health (paper I)
- changes in gross motor function from adolescence to adulthood (paper I)

To provide a deeper understanding of how adults with CP experience

- living with a disability (paper II)
- physiotherapy and physical activity (paper III)
- changes from childhood to adulthood in relation to disability and physiotherapy/physical activity (paper II, III)
- how environmental factors and personal strategies influence everyday life and possibilities for physiotherapy/physical activity (paper II, III)

## METHODS

This thesis consists of two parts: a descriptive study based on quantitative data and an explorative interview study based on qualitative data.

### Subjects

There were two samples in the studies.

The first sample (paper I) was recruited from a cohort of 199 persons  $\geq 20$  years who were identified retrospectively through medical records from the Child and Youth Habilitation in a Swedish county where they had been registered as children and adolescents. The Child and Youth Habilitation in this county care for children with all types of CP diagnoses and all subjects according to GMFCS levels I-V were included in the identified cohort. Based on information from the medical records (from the time the persons were discharged from the Habilitation) sub-diagnoses, motor function, age and gender were noted. With this information 48 people were asked to participate in a clinical examination and structured interviews. The selection was strategic in order to resemble the cohort of 199 people according to gross motor function, gender and age (Table 2). Forty people lived in their own housing (with and without assistance), five in some kind of 'institution', and three with their parents. Three people had children.



**Table 2.** Sample 1. Comparison between the total population (n=199) and the target population (n=48) in relation to GMFCS levels, gender and age. All data are from medical records.

	Total population n=199		Target population n=48	
	n	%	n	%
<b>GMFCS</b>				
<b>I</b>	61	31	15	31
<b>II</b>	48	24	11	23
<b>III</b>	26	13	8	17
<b>IV</b>	32	16	7	15
<b>V</b>	32	16	7	15
<b>Gender</b>				
Women	97	49	25	52
Men	102	51	23	48
<b>Mean age (sd)</b>	32 (8)		33 (8)	

The second sample (paper II and III) was recruited from five counties in the middle and southern part of Sweden. Adults with cerebral palsy who were 35 years or older with GMFCS levels II-IV were included. With help from rehabilitation professionals who had knowledge of the population, 22 people who were 35- 68 years were selected (Table 3). Seventeen people had a daily occupation, whereof two were on sick leave. Four participants were pensioned (three because of disability and one because of age) and one was searching for work. All the participants were living in their own housing and six people received personal assistance in varying extent. Six of the participants had children.

**Table 3.** Sample 2. Characteristics of the adults with CP (n=22).

GMFCS	Population n=22	Men n=12	Women n=10	Mean age (sd) 47 years	Daily occupation n=17
<b>II</b>	7	5	2	42 (8)	7
<b>III</b>	7	3	4	48 (7)	6
<b>IV</b>	8	4	4	49 (11)	4

## Procedure

### Sample I (paper I)

The selected people were invited to participate and were contacted by telephone. During the phone conversation the participants were explained the aim of the study, which gave possibilities to questions and discussion. They were also asked to give their approval to participate. The structured questions and clinical assessments were completed at one occasion by the same physiotherapist (KS) in a rehabilitation department. Each occasion lasted about 2-2.5 hours. Eight of the interviews were proxy reports due to cognitive limitations. The persons who accompanied the individuals with CP were closely related; the proxy was either a parent or an assistant who knew the person very well.

This descriptive study uses information from medical records (see under 'subjects'), structured questions, and clinical measurements/examinations. Questions were asked about living conditions (family, personal assistance, education and daily occupation) and use of health-care, physiotherapy/physical activity (see paper I).

### *Measurements*

#### *Gross Motor Function*

The 6-12 year age band of Gross Motor Function Classification System (GMFCS)<sup>6</sup> was used to classify gross motor function on two occasions: retrospective from medical records representing the period when they were discharged from the Child and Youth Habilitation and at the current clinical investigations (GMFCS is described in the introduction).

#### *Basic Daily Activities*

The physical domain of Functional Independence Measure (FIM; Swedish version)<sup>73</sup> was used to assess the subject's dependence with respect to basic activities of daily living (ADL) such as self-care, sphincter control, mobility indoors and locomotion. The physical domain consists of 13 items with a 7-level ordinal scale, with ratings from total dependence (1) to complete independence (7). The range of the summarized score is 13-91.

### *Instrumental Daily Activities*

The Instrumental Activity Measure (IAM) was used as a complement to FIM in subjects with limited or no need of personal help (the ratings 'supervision' to 'complete independence' according to FIM).<sup>74,75</sup> IAM measures activity levels for people who live in their own housing (mobility outdoors, simple meal, cooking, public transportation, shopping, cleaning, and washing).

### *Locomotion*

The Swedish version of the 13-item Clinical Outcome Variable Scale (S-COVIS), was used to evaluate different aspects of mobility, with emphasis on locomotion.<sup>76</sup> A seven-level ordinal scale for each item ranges from inability to perform a task (1) to normal function (7). The range of the summarized score is 13-91.

### *Passive Range of Motion (PROM)*

PROM was estimated (in all directions in extremities) with a scale graded from 0-2 (0= a few degrees PROM, 1= limited PROM, 2= normal/almost normal PROM). The scale is a part of a motor assessment for stroke.<sup>77</sup>

### *Pain*

Pain intensity was rated on a visual analogue scale (VAS) from 0 (no pain) to 100 (worst imaginable pain).<sup>78</sup> Pain location was described by means of given alternatives.

### *Health-related Quality of Life*

EuroQol (EQ-5D) questionnaire was used for screening health status and self-perceived health in order to get a descriptive profile of the population.<sup>79</sup> The health status consists of five items: mobility, self-care, usual activities, pain/discomfort, and anxiety/depression. For each item there are three levels (1= no problems, 2= some problems, 3= extreme problems). It has an index range from -0.59 to 1, where 1 represents full health. Self-perceived health was estimated using a visual analogue scale where the person indicates the level of his/her own health (0= worst imaginable health to 100= best imaginable health).

## **Sample II (paper II and III)**

The people were contacted by professionals with knowledge of the population in their county or district. They gave short information of the purpose of the study and, if the person permitted, a letter was sent with further information. Next, the subjects were contacted by telephone by the interviewer (KS) and they also gave written consent for the interview to be taped and transcribed.

The interviews took place in the informants' home (n=19) or in a rehabilitation setting (n=3) and each interview lasted between 45-90 minutes.

A qualitative interview study with open-ended questions was carried out to provide insight into how people understand their experiences. The interview contained conversations about living with a disability and experiences with physiotherapy/physical activity. These topics were divided into different areas that should be covered and a general interview guide approach was used giving the interviewer the freedom to have an informal conversation with the informant.<sup>80, 81</sup> Focus was on common daily life situations, but the informants were also asked to describe their experiences in relation to past, present and future concerns. To clarify answers the interviewer asked follow-up questions, rephrased questions and probed for additional examples.<sup>80, 81</sup>

## Statistics and analyses

### Paper I

Descriptive statistics included scores, frequencies, medians, means, standard deviations and ranges for the assessed variables. The Mann-Whitney *U*-test or Student's *t*-test was used for between group comparisons (see paper I). The Wilcoxon signed-rank test was used for within group comparison and Spearman's rank correlation coefficient was used for correlation analysis. The correlation coefficients were valued according to Landis and Koch (Table 4).<sup>82</sup>

*Table 4. Interpreting the correlation coefficients.*

<i>Correlation coefficient</i>	<i>Strength of agreement</i>
<0.00	Poor
0.00-0.20	Slight
0.21-0.40	Fair
0.41-0.60	Moderate
0.61-0.80	Substantial
0.81-1.00	Almost perfect

### Paper II

The analysis in this study was performed according to the Empirical Phenomenological Psychological method (EPP).<sup>83</sup> This method is based upon phenomenological philosophy, which is concerned with the discovery of people's daily life experiences. Five analytic steps were used, designed to

condense and identify themes that reflected the informants' experiences.<sup>83</sup> The first step was reading the transcribed material for each interview. In the second step the transcribed data was divided into meaning-units (MU), discernable parts dealing with the aims of the study, describing different meanings of living with a disability. In the third step, each MU was interpreted and transformed to the researcher's language and understood in light of the whole interview. In the fourth step, each interview was synthesized and presented in a form of synopsis. In the fifth step, the 22 synopses were compared to find meaning structures that were found in all interviews. In this phase repeated reading of the original protocols and the meaning-units gave valuable information. The results obtained from the analysis can be presented as a 'general structure' (if all interviews contains the same meaning structure), or in terms of typological structures or themes, to preserve individual variations. The findings obtained from the analysis in the fifth step in current interviews are presented in different themes.

### **Paper III**

This part of the interviews, addressing physical activity/physiotherapy, was analysed with a qualitative content analysis, a systematic coding of the text resulting in themes that can be seen as an expression of the latent content of the text.<sup>84-86</sup> After selecting the unit of analysis (in this text physical activity/physiotherapy) meaning units were derived from the text by reading the text word for word. A meaning unit is words and sentences from the text containing aspects that are related to each other. Then, the different meaning units were condensed (shortened) and reformulated in other words with the central meaning core preserved. The condensed meaning units were coded with labels representing different meanings of the main topic for this research and the labelled meaning units were grouped in categories.<sup>85,86</sup> The last step was to gain an understanding of the underlying meaning and to find relevant themes with a transition from manifest to latent content.<sup>84, 85</sup>

### **Validity (paper II and III)**

As a validation, the preliminary result was presented to nine of the interviewed people. Their feedback indicated that the interpretation of the interview texts agreed with their narratives. A physiotherapist experienced in rehabilitation and the second author (in paper III) read the interviews. They compared their perceptions of the content in the interviews to the analysis (made by KS) and different interpretations were compared and developed.

## **Ethical considerations**

The first study was initiated and ordered by the County Council of Östergötland. Therefore, no ethical approval from an ethical committee was carried through; however the law of personal records (1998:2004) was applied. A lawyer from the county council and the habilitation chiefs approved the use of medical records. The interview study was approved by the faculty of arts committee for research ethics at Linköping University.

There are some fundamental ethical principles concerning relations between individuals, such as respect for the individual, not inflicting harm, equity and the benefit-risk principle.<sup>87</sup> These concerns are taken in consideration in this thesis. The selection of both samples was strategic and there was a personal relation with the investigator/interviewer within the meetings, which lasted from 45-90 minutes (interview study) to 2-2.5 hours (clinical investigation). Those conditions made it extra important to inform the investigated and interviewed people that they could say no to participate and they were also informed that they could withdraw at any time. However, the researcher (KS) had no therapeutic relation with the investigated and interviewed people, but they were also told that they could contact the researcher again if the meeting had evoked new questions.

## RESULTS

### Gross Motor Function Classification System (GMFCS) and relations to instruments measuring health and functioning (paper I)

There was an almost perfect relationship between GMFCS levels and FIM and S-COVS. There was also an almost perfect relationship between GMFCS and *health status* in EQ-5D and a fair relationship to *self-perceived health* (Table 5). This means that GMFCS have a strong relation to instruments with activity components including gross motor performance and to health-status defined in terms of abilities, but a weaker relation to self-perceived health.

*Table 5. Correlations between GMFCS and FIM, S-COVS, Health status (EQ-5D), Self-perceived health (EQ-5D).*

	<i>FIM</i>	<i>S-COVS</i>	<i>Health-status (EQ-5D)</i>	<i>Self-perceived health (EQ-5D)</i>
<i>GMFCS</i>	r= -0.85 almost perfect	r= -0.90 almost perfect	r= -0.84 almost perfect	r= -0.30 fair

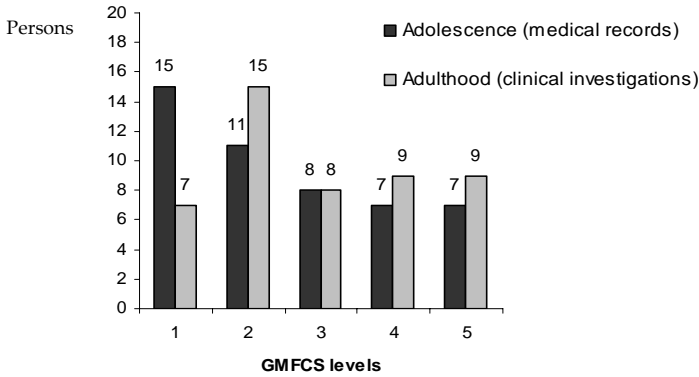
### Descriptions of functioning and health (paper I)

#### Changes in gross motor function from adolescence to adulthood

A comparison between GMFCS levels determined retrospectively from medical records with current classification (clinical investigation, n=48), showed that 16 people had deteriorated motor function in terms of locomotion. There was deterioration in all GMFCS levels (except level V) and there was predominance in level I (Figure I). Three people had changed function in two levels, one from level I to III and two from level II to IV. None had changed to a better functional level.

People with deteriorated function had a significantly higher mean age (37 years) than the unchanged group (31 years). The mean elapsed time since the

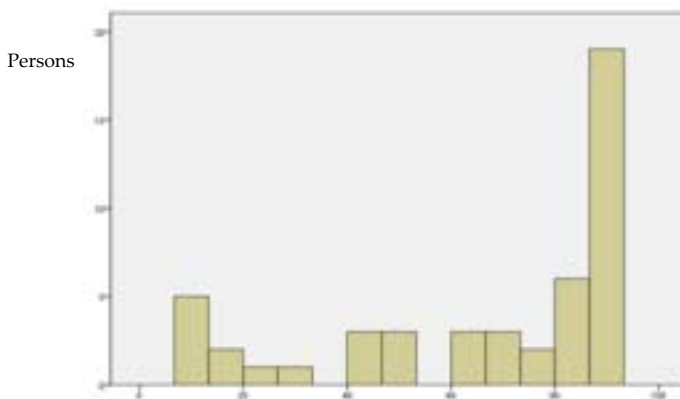
persons had been discharged from the Child and Youth Habilitation was 13 years, and the minimum age for leaving the Habilitation was 15 years.



*Figure 1. Changes in motor function according to GMFCS levels (n=48) from adolescence to adulthood.*

### Activities of daily living and locomotion

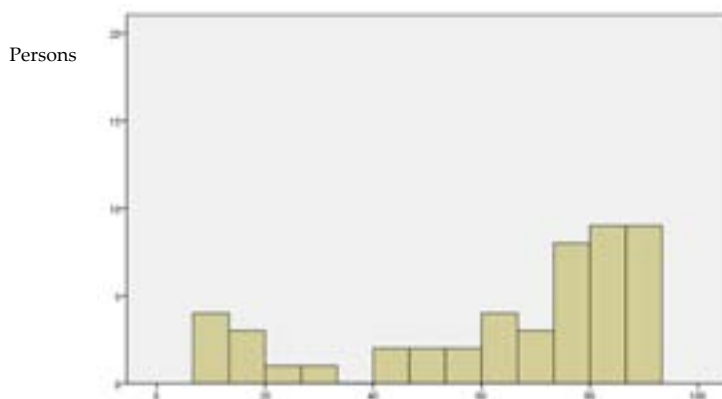
The FIM scores showed a wide distribution in the ability to perform basic personal ADL skewed towards higher scores (Figure 2). The median score was 81. Thirty-four people had no or limited need of personal assistance (complete independence to supervision) according to FIM. Those 34 individuals were also measured by IAM showing lower median scores (3-4) in large-scale shopping, cleaning, washing and public transports, suggesting that these people needed personal assistance in these activities.



*Figure 2. Distribution of FIM scores (physical dimensions). Minimal score 13, maximal score 91 (n=48).*



Locomotion measured by S-COVS had the same profile as FIM (Figure 3), although a lower median score (77). The lowest median scores in S-COVS were the gait variables (such as endurance, assistance and walk distance) and rising from floor, which is illustrated in the figure showing a less skewed figure for S-COVS than FIM.



*Figure 3. Distribution of S-COVS scores.  
Minimal score 13, maximal score 91 (n=48).*

## Passive range of motion (PROM) and pain

Limitations in PROM (estimated as 0-1 in the scale from 0-2) were most common in GMFCS levels IV and V (Table 6). Forty-five people had limited PROM in at least one side and direction. Three people with dyskinesia (GMFCS II) had no limitations. The limitations were most frequent in the ankle joint, including persons with all GMFCS levels.

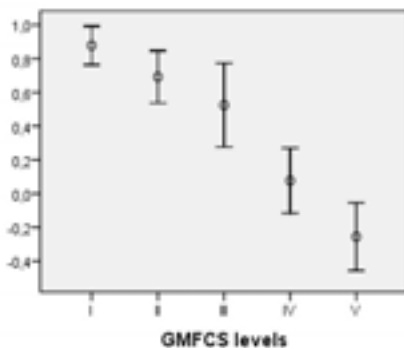
Thirty-three people had musculoskeletal pain in at least one location (Table 6) and there was no correlation ( $r=0.003$ ) between presence of pain and GMFCS levels. The intensity varied from occasions when pain was at its best (mean 8, sd 14) to when it was at its worst (mean 77, sd 19).

**Table 6.** Limited PROM, presence of pain and pain location in relation to GMFCS levels (n=48).

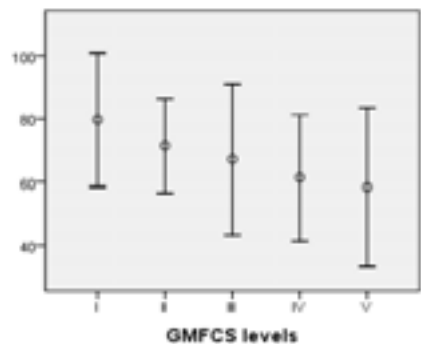
	Total n=48	GMFCS I n=7	GMFCS II n=15	GMFCS III n=8	GMFCS IV n=9	GMFCS V n=9
<b>Limited PROM in at least one direction and one side</b>						
Shoulder	29	2	6	5	8	8
Elbow	11	0	2	2	3	4
Wrist	18	0	5	2	4	7
Hip	29	0	5	6	9	9
Knee	20	0	2	4	8	6
Ankle	39	6	11	6	7	9
<b>Pain in one or more location</b>						
	33	3	12	7	6	5
<b>Pain location</b>						
Neck	10	1	4	0	4	1
Upper extremity	12	1	4	3	4	0
Back	17	3	6	3	2	3
Lower extremity	22	2	6	6	4	4

## Health related quality of life

*Health status* (in EQ-5D) showed problems in foremost the dimensions 'mobility' and 'pain/discomfort' (see paper I). The weighted index was 0.39 (sd= 0.44). The mean score on the visual analogue scale for *self-perceived health* (in EQ-5D) was 68 (sd= 22). Figure 4 and 5 shows the relations of those dimensions in relation to GMFCS levels.



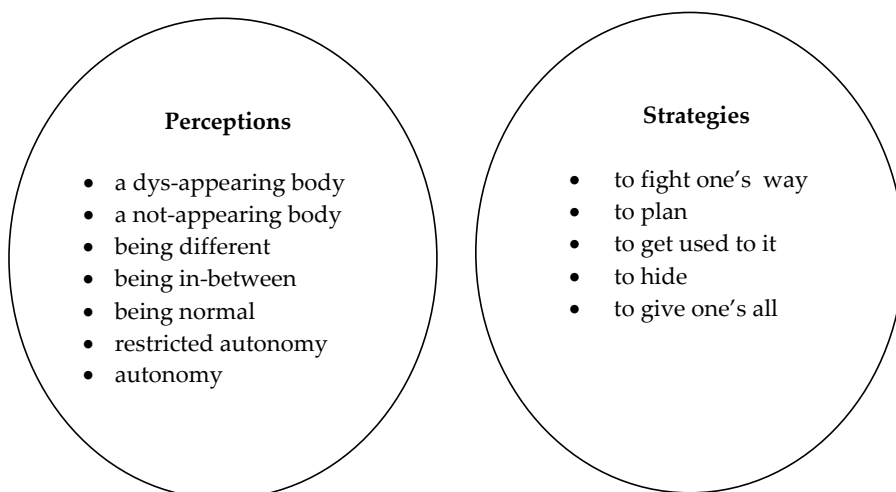
**Figure 4.** Relations between GMFCS levels and health status in EQ-5D (mean, sd) n=48.



**Figure 5.** Relations between GMFCS levels and self-perceived health in EQ-5D (mean, sd) n=45.

## The lived body- experiences of living with a disability (paper II)

The 22 interviewed people, with functional levels II-IV according to GMFCS, had varied experiences in living with a disabled body. Those lived experiences represent the people's interpretation and valuation on life situation. In everyday life, the lived body is essential as we are our body in feelings and actions, but we also have a body we can observe and talk about. The result illustrates feelings (named perceptions) and actions (named strategies) as they are observed and talked about by the informants (Figure 6). It also describes perspectives of change.



*Figure 6. Themes illustrating perceptions and strategies as experienced from adults with CP*

### Perceptions

#### *A 'dys-appearing' body – A 'not-appearing' body*

A 'dys-appearing' body represents feelings of a body that appears in an unpleasant way. It doesn't have to interfere with functioning, but the body is not in the background and there is an awareness of an uncomfortable body, pulling attention from other thoughts and activities. These perceptions were often described by the informants. Musculoskeletal pain, stiffness and fatigue were common 'dys-appearing' sensations. This overriding focus could also be

in situations when the physical defects were highlighted or when the body was unpredictable, as with excessive movements. An often-perceived contributing factor to those sensations was the fast pace of society, as they had to exert themselves to their physical limits. The perceptions of 'dys-appearing' were not new experiences, but had often increased or become more evident over the years. They often increased and led to a dysfunction, but they were often experienced as a slow and imperceptible progress. Eventually, they realized that there had been a change as it interfered with daily activities. When the body was not able to perform the same activities as before, it was evident that 'something had happened' and questions about how to act on this were raised. This also evoked questions about the impact from CP on ageing. On the other hand, there were also perceptions of a 'not-appearing' body: when the body was in background and when they could act without being concerned with the body. For some informants that was the normal condition and for others that could be only for certain periods or in certain situations when other states overshadowed unpleasant body feelings.

### ***Being different – Being in-between – Being normal***

Activity limitations with problems in following other people's pace and doing the same activities had always been a source of perceptions of difference. Feelings of difference were also related to other person's attitudes or treatment. It could happen in varying situations and could be expressed in various ways, such as exaggerated kindness, negative attitudes or bad treatment. Some informants conveyed that being born with a disability means that the disability is integrated in self-image of normality, but changes, including deterioration, created feelings of difference. Beginning to use a wheelchair could, for example, be an evident cursor for difference as the person in wheelchair discovered that he/she was treated in another way than when he/she was walking. Informants that always had been wheelchair users did not have the same experiences, as there were no changes making them aware of that. Self-image is not a static condition. Sometimes acknowledging the disability and in other situations trying to be as 'normal' as possible also created feelings of 'being in-between'. Being affiliated to the CP-group could be experienced as a security and contribute to a positive self-image, but it could also be a stigmatization. In other contexts being just like 'everyone else' could be very important. Positive attitudes and support from friends and family together with meaningful daily activities were stressed as important factors for feelings of normality.

### ***Restricted autonomy – Autonomy***

Autonomy, the ability to control and steer your own daily life, was influenced in a negative way when other people, such as assistants or relatives, decided or when daily life activities were restricted by environmental obstacles. Functional changes leading to decreased activity level also raised questions about autonomy. Individuals who had been independent had to take help from friends and relatives and they couldn't be as flexible in daily life as before, creating perceptions of loss of autonomy. Issues about autonomy were thus not always connected to physical dependence, but to feelings including loss of self-control and possibilities in making one's own decisions. A flexible support had influence on autonomy and some informants were frustrated as society often has a rigid system for support in terms of schedules and accessibility. There were also different opinions whether personal support facilitated autonomy or not. Some informants experienced limited autonomy having another person in their life and never being able to be alone, which also had an impact on personal integrity.

### **Strategies**

The strategies that were emphasized by the informants are common daily strategies, recognizable in all people; however, depending on the situation it could be a positive or negative influence. The strategies listed below are linked to some typical consequences that were stressed by the informants.

#### ***To fight one's way***

Striving for normality and autonomy in being competent and having control over daily situation often resulted in a fight and a determination to manage. This could have a positive impact on self-image, but they were often using their upper limits, which had an impact on body functions with increased pain, stiffness and fatigue.

#### ***To plan***

Planning daily life was a strategy used to meet environmental challenges and cope to time-consuming activities. Planning often resulted in prioritizing that required dropping certain activities, in order to manage work and family.

#### ***To get used to it***

Daily life situations with setbacks combined with a 'dys-appearing' body and deterioration in function often resulted in an acceptance of life situation and

deterioration. A consequence of this could be an inability to control or to initiate certain activities.

### *To hide*

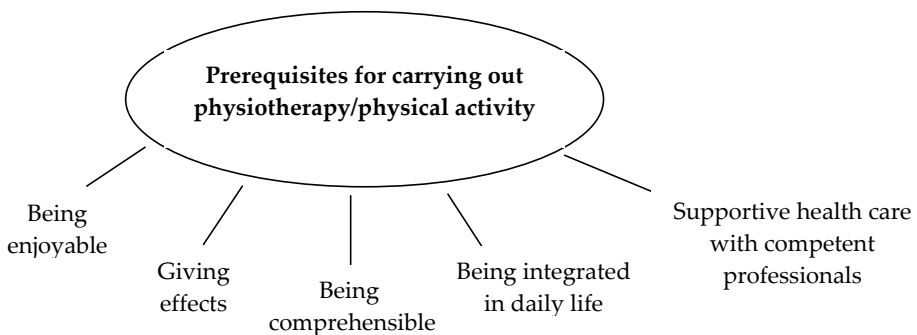
Avoiding situations where the disability is prominent and keeping distance from others were typical hiding strategies. This was a way to create a better self-image, but could also result in an own stigmatization of the disability as stressing the difference by hiding a 'part of your self'.

### *To give one's all*

Some informants stressed the importance of paying attention to non-disabled people and their uncertainty towards disability. This is a prerequisite for creating a positive atmosphere, which often ended in increased self-confidence for all people involved in the interaction.

## **Prerequisites for carrying out physiotherapy and physical activity (paper III)**

The narratives from the 22 interviewed people illustrated a variety of experiences from childhood to adulthood. These experiences revealed important prerequisites for carrying out physiotherapy/ physical activity. They are summarized in five themes (Figure 7).



*Figure 7. Prerequisites for carrying out physiotherapy/ physical activity.*

## **Being enjoyable**

Leisure activities, when combining training and own interests are enjoyable as they also focus on the activity and not on the body. Although experiences differed, as children many informants experienced physiotherapy as a duty imposed by authorities consisting of experts and parents. They experienced monotony, pain and stretching highlighting a dysfunctional body. Similarly, adulthood training can be experienced as a duty, but adults can exercise some sort of autonomy by choosing to participate in activities they enjoy, although it is not always so easy to find those activities

## **Giving effects**

Obvious activity improvements were regarded as effects; however there was an uncertainty about how physical activity contributed to those effects and different thoughts about advantages with intensive training or using daily activities. Some informants thought that in a longer perspective the previous days physiotherapy had contributed their present condition. An often-mentioned positive effect was the relationship between physical activity and well-being, described as reduced pain, fatigue, and stiffness.

## **Being comprehensible**

As a child it was often difficult to understand the meaning with physiotherapy/physical activity and the explanations were often in relation to the future, which was abstract and uninteresting. This often resulted in that they finished physiotherapy in adolescence when they could make their own decisions. In adulthood there is an underlying comprehension about the meaning of physical activity, but the informants also had many questions about physical activity in relation to dysfunction and ageing.

## **Being integrated in daily life**

To integrate physical activity in daily life was regarded as important, but it could be a challenge. Time-consuming daily living, transport problems, limited accessibility and/or lack of personal assistance were common obstacles. As an adult those barriers were even more evident, although some informants could capture the opportunities using daily activities as training.

## **Supportive health care with competent professionals**

The importance of support from health care and competence from professionals was unanimous and expressed as limited or absent as an adult.

The informants described problems in finding where to turn when there was a need for support and treatment related to the disability. There were many experiences of unprofessional treatment with limited sensitivity and knowledge about CP. The informants stressed that there must be a structured follow-up from health care and physiotherapists with knowledge of CP.



## DISCUSSION

### Methodological considerations

Classification according to GMFCS was made at the individual meetings with the adults with CP. The experience from current study is that GMFCS was an easy administered tool. The GMFCS had strong correlations to FIM, S-COVS and EQ-health status, which indicate that GMFCS for adults seems valid. GMFCS was also used to classify according to medical records. GMFCS has earlier been used based on retrospective chart reviews and this was considered to have good inter-rater and test-retest reliability.<sup>88</sup> Adults with CP have also rated themselves according to GMFCS and this was compared to classifications done by a physiotherapist.<sup>7</sup> The correlation showed excellent agreement. This indicates that GMFCS can be used in the adult population and that retrospective rating according to written journals is appropriate. The new version of the GMFCS, expanded to include age range between 13-18, can now replace the 6-12 age range when classifying gross motor function in adults.<sup>8</sup>

In the first study (paper I) there was a strategic selection in order to have the same GMFCS levels, mean age and age distribution as in the cohort of the identified 199 individuals. The GMFCS levels based on medical records had similar distributions as in other surveys,<sup>11, 89, 90</sup> although those surveys showed higher percents in GMFCS I and II. This difference could be because some adolescence with minor disabilities had been discharged or perhaps never enrolled in the Habilitation. Sub-diagnoses of CP were noted, but information about cognition was insufficient in the medical records in order to classify cognitive levels. However, the base for this study was gross motor function and therefore aspects of cognition and sub diagnoses were limited, which also can be regarded as a weakness in this study.

Paper I included both clinical investigations and self-reported measures. The assessments were carried out by one physiotherapist. The structured assessments (FIM, IAM and S-COVS) were easy to administer and the physiotherapist had previous practise in using these instruments. In the assessments of passive range of motion (PROM) the main extremity joints were measured in all directions. The scale from 0-2 can be questioned, but

measuring with a goniometer was considered too time consuming. As only frequency and location of decreased PROM has been used in the result, the used estimation scale was appropriate. Of the subjective ratings of pain and health some were proxy reports and these were used in the results. The people who accompanied the subjects had good insight into the assessed persons' functioning and health in a longer perspective, although three of the accompanied persons did not estimate self-perceived scale in EQ-5D. Proxy evaluation could have influenced the results as there can be both over- and under estimations especially according to pain and self-perceived health.

The statistical analyses (paper I) were mainly descriptive. Forty-eight people are a small sample and mainly non-parametric statistics were used and sometimes the information is presented only by visual information in a figure or a table. Therefore, there must be a caution when drawing conclusions given by statistical means.

In the interview studies (paper II and III) there was also a strategic selection, but the selection was narrower according to GMFCS levels (II-IV). Because the subjects were selected through personal contacts sub-diagnoses were not available through medical records. Cognitive level was also not used as a parameter and all informants had cognitive abilities that allowed them to participate in conversation.

The interviews were carried out by a physiotherapist, who had special knowledge about the topics in the interviews, which can be both an advantage and disadvantage. The cause of validity is often discussed in relation to qualitative studies and concepts such as credibility or trustworthiness are also used.<sup>91</sup> This study may be limited by the fact that the interviewer also conducted main part of the analysis. However, another physiotherapist read the interviews, compared to the preliminary analysis and provided feedback. The second author in paper III took part in the last steps of the analysis of physical activity/physiotherapy. This is one way of validation, an is also called 'peer debriefing'.<sup>91</sup> The preliminary results were presented to nine of the informants; they provided feedback and confirmed that the results agreed with their narratives, which can be regarded as a 'member checking'<sup>91</sup> or communicative validity.<sup>80</sup>

Validity also depends on the clarity of the steps in the analysis and how this is described, as this sorting procedure means that the text is lifted from its

context.<sup>92</sup> The study's main aims - experiences of living with a disability and physiotherapy/physical activity - resulted in two complementary analyses. Both according to the 'empirical phenomenological psychological method' (EPP) and the 'qualitative content analysis' the steps in the analyses were distinct, but there were differences. In the EPP-method one interview was analysed as a whole, except in the last step. In the qualitative content analysis there was a de-contextualizing at the beginning of the analysis, which was regarded advantageous as the first analysis with EPP had provided a wholeness of the underlying content in the interviews.

Generalisation is an ongoing process in daily life, but according to scientific studies there are certain criteria that have to be fulfilled in order to generalise. There are different opinions whether qualitative material can be generalised.<sup>80,91</sup> If there is a generalisation, its criteria should be carefully scrutinized.<sup>93</sup> The first question is about the population and how representative it is for the studied phenomena.<sup>93</sup> In this study, the population is selected to represent a certain subculture. Because this culture also must be looked at from different perspectives,<sup>93</sup> the variables, -as motor function, gender and age- were selected to include many perspectives. The theoretical deduction in this study relies on assumptions that there are some fundamental conditions that are shared in this sample as the interviewed people are from a special context in terms of culture and the health care system. Another discussed issue according to generalisation is the size of the studied population. In qualitative studies, there is a problem if you want to draw conclusions from a small sample; however a large sample also presents problems in doing thoroughly interpretations.<sup>80,92</sup> In this study there was a rather large sample, but the sample was not so large as to make interpretations difficult. In sum, according to the representation of a certain subculture and the relatively large sample size we conclude that there are possibilities to make some generalizations from these interviews.

## Discussion of results

An important message conveyed during the personal meetings in both clinical investigations and interviews, is that persons with CP not only are heterogeneous according to functioning, but also according to experiences and thoughts about living with a disability. This is important as many of the interviewed people thought that they were viewed as a 'CP-collective' instead of being seen as individual people. In the following discussion certain topics

are illuminated and therefore it is important to have in remembrance that these topics not always are general conclusions, but important issues stressed in the discussion. Furthermore, in the light of selection and process of change in the population, there is also not a major focus on people with very small restrictions or on those with severe disabilities in the following discussion.

## **Deterioration in gross motor function according to GMFCS**

One third of the 48 individuals (paper I) had deteriorated according to GMFCS levels. Also other studies describe deteriorated gross motor function in approximately one third of their samples.<sup>7, 34-37</sup> In the current study, above all, individuals in level I had changed to level II. This can be a source of error, as it can be difficult to make explicit distinctions between level I and II and that the first rating was retrospectively assigned medical records. On the other hand, the investigated people confirmed that there had been a change in gait performance since their adolescence. On account of the use of GMFCS, no deterioration from level V could be registered, although there probably were changes in body functions.

Normal ageing means decline in functioning, but the mean age of the individuals who had deteriorated in gross motor function was 37 years (paper I). Although this finding should be taken with caution as the number is based on only the 16 individuals who had deteriorated, other studies confirm that this deterioration often happens earlier in people with CP.<sup>35,37</sup> There are, however, some contradictory results. A study concerning stability of the Gross Motor Function Classification System in adults, established that GMFCS levels observed at the age of 12 years is highly predictive for adult motor function.<sup>94</sup> The mean age for the participating adults was 22 years and perhaps the same study in an older age group had given another result. There are also different definitions about what should be considered as a decline in motor function in relation to one's own choices and environmental restrictions.<sup>95</sup> This can also contribute to different conclusions

## **The 'dys-appearing' body and its impact on daily activities and self-perceived health**

The concept 'dys-appearing' body was created by the philosopher Drew Leder.<sup>96</sup> He uses it when describing a body that appears in an unpleasant way and it is not always equal to 'dysfunction'. The body 'stands in the way' and attention is pulled back from other activities. This concept was found

appropriate and was used as a label to describe the informants' narratives (paper II) about their feelings of pain, stiffness and fatigue.

Musculoskeletal pain is an often described problem in persons living with CP.<sup>27, 29, 31, 32, 97</sup> The informants in current interviews (paper II) also described that pain often had followed from childhood into adulthood with many changes in intensity and locations. The investigation (paper I) showed that pain was common in all GMFCS levels and there was no correlation between pain and functional levels. The most common location was back and lower extremities locations that are also described in other studies<sup>27, 29, 31, 32, 97, 98</sup> and indicates that pain can be related to walking ability.

Musculoskeletal pain is often connected to repetitive strain injury, which in the CP population is related to greater efforts at mobility.<sup>29,31,97</sup> The investigation of the 48 individuals (paper I) showed that it was a large divergence in persons when pain was 'at its worst' and 'at its best', which is indicative of load-related pain. People with CP have described activity and exercise, but also rest, as relieving factors.<sup>29,31,98</sup> Studies also report small interference of pain in daily activities.<sup>29,32,97</sup> The informants in the interviews illuminated a double strategy, both trying to neglect pain as long as possible, but also acknowledging pain when it intruded on daily activities, which resulted in reprioritizing and new strategies. A study about coping with chronic pain showed that people with CP are more likely to use persistence in tasks and less likely to use guarding and rest strategies than people with a primary diagnosis of chronic pain.<sup>99</sup> This is interesting information, suggesting an 'overexertion theory', and the importance of finding activities that prevent pain, but still preserve activity levels.

Decreased joint motion according to passive range of motion (PROM) was described in paper I and, as expected, there were more limitations in individuals with major disabilities. The most common limitation in PROM was in the ankle and this was also frequent in people with GMFCS levels I and II. No other study has paid attention to this, but it is also important to realize that this relatively small decrease of joint motion can lead to walking problems and pain and is worthy of further investigation. This objective, measured range of motion (paper I), was identified by the informants in the interviews (paper II) as part of the concept 'stiffness', which contributed to a 'dys-appearing' body also including problems with muscle tone and muscle strength.

In a Norway population, adults with CP reported more physical fatigue, but not more mental fatigue, than in the general population.<sup>30</sup> The same study also showed that fatigue was most prevalent in people with moderate impairments, suggesting that these persons have an imbalance between workload and work capacity in daily life. The informants (paper II) acknowledged that fatigue was a common state and they often related to their struggle in managing daily activities and keeping up with today's tempo as a predominant cause. Another study has also reported that fatigue is associated with low sense of coherence, and people who experience fatigue find daily life more unmanageable and incomprehensible,<sup>100</sup> which implicates that there is a need to gain a deeper knowledge in the reciprocal relation of these conditions.

'Dys-appearing' perceptions did not always interfere with functioning, but gradually it was evident in loss of daily activities. Many informants talked about a slow and imperceptible progress and the conditions were not explicit until it really interfered with daily life. This is surprising, as we know that people with CP, compared to people without CP, have faster onset of body ailments and deterioration.<sup>35,37</sup> Strategies, such as accepting or hiding the problems, can contribute to these feelings of a slow process.

In spite of the above mentioned problems, many individuals (paper I) were almost independent in personal ADL, which gives a picture of a population that manage many basic daily activities. However, motor function showed problems with gait-variables such as endurance, assistance and walk distance; many people with moderate CP had problems with large-scale shopping, cleaning and public transportation. The interviews (paper II) confirm this picture; persons with GMFCS levels II -IV struggle to manage daily life, but pain, stiffness and fatigue negatively impact daily life. Despite struggling, dependence in ADL increased significantly in a group of community-living adults with CP in a perspective of five years<sup>101</sup> and gross motor function is reported as one determinant for restrictions in daily living.<sup>43</sup> These conditions demand significant prioritization, finishing certain activities, which in turn creates a vicious circle with reduced activities.

Pain and fatigue are factors that are reported to have negative impact on well being and quality of life for people with chronic disabilities.<sup>102, 103</sup> In the current study (paper I), mean score on the visual analogue scale (EQ-5D)<sup>79</sup> for self-perceived health was 68. In the general population, in the same age-group in a Swedish population, mean score was 82.<sup>104</sup> The lower mean score in people with CP can have different explanations, such as pain and deterioration, but

other aspects concerning life-situation can also be affected. On the other hand, this study (paper I) and other studies have shown that there was no or minor correlation between self-perceived health and functional level.<sup>38, 47-49, 105, 106</sup> This has been called the 'disability paradox'<sup>103</sup> and shows that functional ability in itself and self-perceived health are not related, but current interviews (paper I) indicate that functional deterioration negatively impacts well-being and self-perceived health.

## Self-image

Perceptions of both difference and 'normality' were often identified in the interviews (paper I). This can be described as the mental picture one has of one's self, often named self-image or identity. The informants' highlighted two main causes for feelings of difference: activity limitations and other people's attitudes and treatment.

Activity limitations were often discussed in relation to time; many activities were time consuming and they compared themselves with other peoples' fast pace. This created frustration not only because the elucidating of difference, but also as it interfered with daily life. These limitations were also related to changes, especially deterioration. Many of the informants noted that because they were born with CP, this is their 'normality'; however new circumstances made them reconstruct identity and individuals with mild CP suddenly felt disabled. An acquired disability in adulthood is sometimes called a biographical disruption, an awareness of a distinct difference between past and present.<sup>107</sup> A person with a congenital disability has no distinct cut-off, but still there is a biographical change that depends on the past and of the expected future, and this also influences self- image.

Other people's attitudes and treatment were described and discussed by the informants as important cursors for perceptions of difference. This could happen in many situations, including when meeting professionals in social- and health care. Toombs<sup>108</sup> discusses the experiences of being looked at in the meeting with health professionals and notes that the body becomes an object in the 'gaze of the other'. Another described aspect is that when we make people 'other', we group them together as objects and diagnosis is prior to the individual.<sup>109</sup> People with CP have long experiences of being investigated and valued and the informants had varying experiences and opinions about this.

Another dimension concerning a 'dys-appearing' body in relation to others attitudes was the perception of an unpredictable body with uncoordinated and involuntary movements, which often increased with different emotions. This was described as a loss of privacy and sometimes resulted in hiding strategies by avoiding certain situations. This is discussed by the sociologist Goffman<sup>110</sup> in the perspective of the 'gaze of the other'. He talks about 'giving' information as our attempts to control our bodies as we want others to perceive our body. On the other hand, our body will always give more information than we can control, which he calls 'give off' information. For persons with CP, this 'giving off' often is harder to control and gives other people access to their thoughts and feelings.<sup>108</sup> Except for these body expressions, body appearance was not a significant issue in the interviews. Some of the informants talked about uncertainty about their appearance especially in adolescence, but perhaps in this group of adults, and ageing people, other things were more important than physical appearance. This has also been confirmed in other studies about body image in people living with longstanding physical disabilities<sup>111,112</sup> and it is also reported that physical appearance had less impact on quality of life than performance of daily activities, which is expressed as a 'can do' approach to life.<sup>103</sup>

One common theme was thoughts about using a wheelchair in relation to self-image according to others' attitudes and treatment. Informants who had been walking and had begun to use a wheelchair had many opinions about pro and cons. They felt they were treated worse when they used a wheelchair, but those who always had been using a wheelchair did not have the same feelings, showing that a usual condition from childhood is 'normal' for that person. The upright posture has been discussed in different philosophical reflections, arguing that the upright posture is the constitution of lived spatiality and that vertically is related to autonomy.<sup>108</sup> A sitting position also causes others to assign a dependent role to the sitting person and attitudes toward an individual in sitting position can be difficult to change. Professionals in rehabilitation have to deal with questions about using a wheelchair or not. We often recommend a wheelchair in order to reduce strain and increase activity, but as this also is a question with existential dimensions, with the wheelchair as a cursor for disability, we need to support the wheelchair user in their coping strategies. Training children to walk to the limits of their ability can be questioned when they have to use a wheelchair as adults. There are however opinions from adults with CP that such an approach was worthwhile, even if as adults they become wheelchair users.<sup>35, 113</sup>



Another matter of self-image is that some informants had perceptions of living 'in-between', sometimes being affiliated to the disability group and sometimes being seen as a 'normal' individuals. It is described how young individuals with disabilities identified themselves as 'part of two worlds'.<sup>22</sup> This ambiguity has also been discussed in relation to transition from adolescence to adulthood when group affiliation plays an important role in forming identity.<sup>114</sup> Which group am I affiliated to, to the disabled or the other friends in my age? However, parents and close related friends importance for a positive self-image were stressed in the interviews (paper II) and similar results is shown by others.<sup>115-117</sup> It is suggested that self-image is formed early in life and (making the assumption that parents in general have positive attitudes towards their child) when social interaction outside the family do occur this can lead to a more negative self-image with feelings of being different.<sup>115</sup>

### **Being autonomous**

Autonomy is not necessarily connected to physical independence, but to the ability to control and steer one's own daily life. Hindering factors, which could be both environmental and personal, occurred when they intruded on decisions and personal privacy. Personal assistants could, for example, simultaneously promote and hinder autonomy, also discussed in another Swedish study.<sup>117</sup> A hindering factor could be that personal assistants were present even in personal situations and that their daily schedule sometimes determined over the individual. A study also showed that individuals who needed more mechanical assistance had higher self-efficacy than individuals with personal assistance.<sup>118</sup> Self-efficacy is nearly related to autonomy as it deals with decision making,<sup>119</sup> and this shows that decision making is more complicated in relation to personal dependence than dependence of mechanical assistance.

Informants, who had deteriorated in function and lost certain activities, often highlighted these losses in relation to autonomy. Deterioration thus seemed to be an important factor for perceptions of restricted autonomy. The first step was often to ask closely related persons when they needed help, which made them feel like burdens. They also noted that environmental barriers influence autonomy. For example, rigid society systems such as health and social care, often intruded on their own decision making about when and how they wanted to do certain activities.

Decisions made by parents and other caregivers in childhood and adolescence were common topics in the interviews, also described in another study; many perceived help from parents as a barrier and overprotection.<sup>117</sup> This is also linked to problems with comprehensibility in adulthood.<sup>100</sup> Being autonomous also depends on individual competence and one's own abilities to make choices. There must be a correlation between our goals and our capacity, and an autonomous individual can reflect upon choices.<sup>120</sup> Clearly, parents and professionals must guide young people to make decisions that control their daily life, as well as help them learn how to make realistic goals. The importance of balancing challenges and personal recourses develops the sense of manageability<sup>100</sup> and is also regarded as a factor for high quality of life.<sup>103</sup>

### **Physical activity and physiotherapy**

The interviews (paper III) gave insight into experiences of physiotherapy and physical activity and one matter of concern was the importance of enjoyment and pleasure as important factors for carrying on in a longer perspective. Memories from childhood were not always enjoyable and other studies have reported the same,<sup>69,121</sup> but we also know that methods have changed since people in this sample were children, and hopefully integration, home-based habilitation and new approaches to physiotherapy have had a positive impact on those experiences. A physiotherapist,<sup>122</sup> who have interviewed younger persons with CP and spina bifida, found that they had similar experiences as older adults, and concluded that the body often is fragmented as physiotherapists often highlights physical defects and focus on impairment, an approach still used by physiotherapists; it is more to produce movements than to experience joy in movements. The informants in present interviews also gave examples of training as enjoyment in childhood when, for example, they could focus on some activity and did not have to focus on body impairments. As adults it is of course also important that physical activities are enjoyable and a new study about adults with CP has also showed that enjoyable activities (such as pool/snooker and bowling) were more correlated to health related quality of life than maintaining and improving fitness and physical function (such as physiotherapy, stretching and gym activities).<sup>70</sup> These enjoyable activities often included social interaction, but on the other hand, they were rather low intensive activities. Therefore, a challenge must be to find activities that are intense enough to affect change or stability, but also enjoyable.

According to effectiveness, some informants were convinced that physiotherapy in childhood had contributed to today's condition and therefore had a positive attitude towards training effects. On the other hand, there was also an uncertainty about the benefits of physical training and some effects were seen as negative, such as causing pain and fatigue. Still there are few studies about effects of different types of physical activities in adults with CP. As mentioned before, studies have shown effects of strength training with impact on daily activities<sup>60-62</sup> but the samples are small and more studies are needed. Although physical activity should also address cardio respiratory, only two studies were found. One of these studies showed that physical work capacity for individuals with ambulatory spastic CP is low compared to adults without disabilities.<sup>123</sup> The same study showed that the subjects were performing close to their physical limits in daily activities. A follow-up to this research resulted in a cardio respiratory fitness program, which gave significant improvements for the adults with CP.<sup>64</sup> As fatigue was an often-mentioned state in current interviews (paper II), activities that improve aerobic power and physical work capacity should be emphasised. It is supposed that adults with CP are not engaging in enough physical activity with sufficient intensity to gain associated health benefits.<sup>70</sup> Today there are guidelines about intensity and time amount for general population and other disability groups,<sup>59</sup> but it can be questioned whether these are applicable to CP.

Another effect of physical activity, often mentioned by the informants (paper III), was well-being, which could be physical, such as reduced pain and stiffness, but also mental. Generally people feel good when they have achieved something and this relation is also indicated by other researchers, who show that physical activity has an impact on quality of life, well-being and body-image for people with disabilities.<sup>124-126 127</sup> These aspects are also important to pay attention to as a physiotherapist and therefore not only physical effects, but also well-being is important to evaluate as a parameter of effects.

The informants (paper III) told that as children they sometimes found it hard to comprehend why they had to do certain physical activities. They did not understand the explanations as they often were in relation to adulthood and as adolescence they were fed up with physical training, resulting in giving up those activities when they could make their own decisions. On the other hand, another study also describes that there is too little explanations and that there is no focus on adult living in childhood.<sup>121</sup> This is now discussed when talking

about transition to adulthood, suggesting adolescence to be more active in decision making<sup>128</sup> and it is also reported that people with CP who as adolescence had learned how to be responsible for their health were more physically active as adults.<sup>69</sup> The difference between congenital and acquired disabilities is also reflected in the context of motivation with the assumption that a person with an acquired disability is more motivated.<sup>69</sup> Now, as adults, the informants (paper III) understand the benefits of physical activity, but they want more support and knowledge about physical activity and ageing issues, in relation to their disability. They also have problems finding support in decision making and in finding appropriate alternatives that would help them be physically active.

The informants wanted ways to integrate physical activity with daily life, a problem that is shared with many adults in today's society. Disability, as such, is a barrier because it is time consuming due to pain, stiffness and fatigue. Some of the informants could use ordinary daily activities, such as using stairs instead of elevator, but that was rare. If they wanted to take part in some organized fitness training, there were many environmental barriers as accessibility, costs and problems with timing according to other activities. These problems are also described in other studies.<sup>67,72,105</sup> Other aspects are that fitness personnel must be more knowledgeable about disability and a stronger relationship must be established between fitness professionals and physiotherapists<sup>129</sup> and more and better trained 'adapted physical activity experts' are needed.<sup>124</sup>

A prerequisite, in order to educate fitness professionals, is that physiotherapists must have knowledge about adulthood and ageing with CP. Unfortunately, the informants' experiences was that, as adults, this is seldom the case when they meet physiotherapists. They experienced problems in finding competent professionals and in getting support and training in a longer perspective. This is a difference to the support provided by the Child and Youth Habilitation and discussed in many reports as the decrease in use of health and rehabilitation services in the transition from adolescence to adulthood.<sup>20,32,71,129-131</sup> There are also studies showing that adults with complex care needs have more admissions to health services than the general population, but there are problems with limited access to specialist physicians.<sup>132, 133</sup> Consequently, problems with access and limited competence create frustration and feelings of being misunderstood and even sometimes offended (paper III). As described (paper II) different causes results in a loss of initiative to 'fight the system', and a vicious cycle starts.

## **Transitions in life and strategies to meet them - implications for professionals**

Living with a lifelong disability means the same transition phases as for everyone else, except the influence of the disability. The interviews showed that strategies in meeting daily life and transitions in life also are universal, but the consequences can have other impacts on life for persons with disabilities. In spite of faster deterioration in body functions than in normal ageing, the process is experienced as slow and imperceptible and the informants also illuminated that deterioration is not only a matter of loss in activities, but also changes according to self-image and autonomy.

Many individuals neglect opportunities to get support and help in for example physical activities, after leaving the care and supervision of the Child and Youth Habilitation. Living with a congenital disability with constant problems, which often are taken for granted, can also influence how long individuals wait to find services. This also addresses an important task for professionals in supporting individuals during this transition phase when decisions about daily living should be taken. Today's discussion is concentrated on the transition from adolescence to adulthood, with respect to personal changes, such as, for example, the acquisition of adult social roles and increased autonomy.<sup>19,134,135</sup> However, an insight from the current interviews is that it is important to meet the individuals between 25-30 years, as it is in this time-period that they begin to reflect over functional changes and want to prevent deteriorations.

Another phase in life, still sparsely documented in research, is the transition into ageing.<sup>136</sup> Strauss et al.<sup>17</sup> describe a marked decline in ambulation in late adulthood and few of the 60 year-olds who walked well preserved this skill over the following 15 years. However, other skills such as speech and self-feeding were well preserved. They also concluded that survival was poorer among those who had lost mobility, but still we need more studies about ageing issues in CP. A report from Ontario indicates that middle adulthood was a difficult time for people with CP, especially with feelings of loneliness and reduced well-being. In older ages, well-being increases again as, for example, pain is reduced.<sup>137</sup> The informants in current interviews had many questions about the impact of disability on ageing and they also expressed feelings of unpredictability and anxiety towards the future.

Today's focus among professionals is on childhood and adolescence, but current interviews indicate that we need a specific competence in also meeting adult and ageing persons with congenital disabilities. King et al.<sup>138</sup> have also shown the importance of understanding the individual needs for different kinds of support as, for example, emotional, instrumental or cognitive support. It is important to incorporate the individual experiences and subjective perceptions of a disabled body in the goals of rehabilitation as 'best possible function'. In rehabilitation settings, independence (or reduced dependence) often is an overriding goal. It has been argued that traditional definition of independence is mechanic and technologic<sup>114,139</sup> and that an important goal must be to empower people with CP to take control over their own health and enhance self-efficacy.<sup>140</sup>

The core in physiotherapy is physical activity and a big task for physiotherapists is to find physical activities that promote well-being and prevent decrease in daily activities and problems with pain, stiffness, and fatigue. Knowledge in how self-image and autonomy are shaped and their relations to physical activity, together with sensitivity in seeing consequences of different coping strategies, is essential for a physiotherapist. Cognitive function is not highlighted in this thesis, but is, of course, important and discussed in other studies, showing the importance in having knowledge about usual problems with for example learning, memory, and perceptual modalities, as well as differences in cognitive profiles in for example spastic and dyskinetic CP.<sup>141, 142</sup>

A challenge is also that we still have limited knowledge about the relation between load-related problems versus positive effects of physical activity. The risks and benefits, described as the relation between physical activity and load, are important to explore.<sup>143</sup> There is a need to evaluate effects of different interventions and their long-term benefits and the importance of better knowledge about cardio respiratory fitness is highlighted<sup>143</sup> as this is stated as an important goal for the general population.<sup>59</sup> It is also argued that future research about physical activity for people with disabilities must include more information on the physical activity profiles of specific subgroups and determine barriers and facilitators.<sup>144</sup> This thesis contributes to these issues.

This thesis showed that there are many structural barriers in society in general and specifically in health care. These are problems with, for example, access, transportation and costs. Physiotherapists are charged with improving

possibilities for adapted physical training and finding possibilities to integrate physical activity into daily life. The informants wanted structured follow-ups and inherent in this is also a need to establish a 'physical place', where the people always can turn to when they need support and treatment from professionals with competence about CP. This points to the importance that rehabilitation professionals also must take part in, and influence, disability policy to improve the social and health care for people with CP.

## CONCLUSIONS

In a CP population representing a spread of functional levels there are deteriorations according to gross motor function in about a third of the population. This occurs in early adulthood and can not be regarded as normal ageing. In spite of deteriorations, a majority manages personal ADL, but there are more apparent problems in instrumental ADL. Problems experienced in relation to deterioration are pain, stiffness, and fatigue. These problems are often experienced as slow and imperceptible and are neglected until they really interfere with daily activities.

A congenital disability is an integrated part of the 'self' and experienced as normality for the individual. Perceptions of difference are often related to limits in taking part in daily activities and/or in other people's attitudes and treatment, but are also influenced by deterioration in functioning. Possibilities in making one's own decisions and control of daily life can be more important than physical independence in itself and become explicit in connection with decreased abilities to manage daily activities.

Experiences of physiotherapy and physical activities vary through life. In childhood, there was often limited comprehension about the meaning of physiotherapy, and many physical activities were experienced as a burden and not enjoyable. In adulthood there is a deeper comprehension about benefits with being physically active, but there are often problems with integrating physical activity in daily life and to find professionals with knowledge of CP.

Physiotherapists must provide lifelong knowledgeable support for people with CP that attends to different life situations and individual strategies. It is also important to provide continuous follow-ups with competent professionals.

Further research and development are needed in order to gain more knowledge about different aspects about living with a lifelong disability in order to improve the conditions for the coming generations of adult and ageing people with CP. Knowing that the majority of the population has a minor motor disability (GMFCS I) it is also important to illuminate adult and ageing perspectives from this subgroup.



## SAMMANFATTNING

Cerebral Pares (CP) är ett livslångt funktionshinder, men trots detta har de studier som publicerats framförallt inriktat sig på barn med CP. På senare år har dock intresset ökat gällande olika aspekter av att leva som vuxen med ett medfött funktionshinder, vilket i sin tur har medfört att nya studier publicerats.

Det övergripande syftet med denna avhandling är att bidra till ökad kunskap och förståelse av att leva med cerebral pares som vuxen med fokus på 'den levda kroppen' och sjukgymnastik/fysisk aktivitet.

Avhandlingen baseras på två populationer. Den första (studie I) består av 48 individer med en variation i motorisk funktion som motsvarar den generella CP-populationen och innefattar alla fem nivåer enligt en grovmotorisk klassifikation (Gross Motor Function Classification System, GMFCS). Den andra populationen (studie II och III), består av 22 personer och representerar individer med motorisk funktionsnivå två till fyra enligt den grovmotoriska klassifikationen (GMFCS).

Studie I innefattar strukturerade frågor samt klinisk undersökning med vedertagna utvärderingsinstrument och analyseras med kvantitativa metoder. Studie II och III baseras på intervjuer och analyseras med kvalitativa metoder.

Det övergripande resultatet visar att det sker en försämring av motorisk funktion för många personer redan i tidigt vuxenliv. Dessa försämringar föregås och/eller följs av muskuloskelettala problem som smärta och nedsatt rörlighet, ofta i kombination med trötthet. Trots försämringar upprätthåller många sina personliga dagliga aktiviteter, men tvingas ofta prioritera bland samhälleliga aktiviteter.

Försämringen upplevs ofta som långsam och smygande, men den medför också att självbilden förändras med nya tankar om annorlundaskap. Detta kan upplevas i relation till aktivitetsinskränkningar, med svårigheter att ta del i andra människors aktiviteter, men också i relation till hur man blir bemött och behandlad av andra människor. Försämringsprocessen påverkar också autonomin, med minskad förmåga att själv kunna styra över sin dagliga tillvaro.

Viktiga förutsättningar för att utföra fysisk aktivitet i ett längre perspektiv är att det ska vara ett nöje, ge effekter, vara förståeligt och kunna integreras i det dagliga livet. Det är också viktigt att få stöd av kompetenta professioner inom hälso- och sjukvård. Erfarenheterna av dessa förutsättningar varierar och framförallt i vuxen ålder är bristen på kompetent stöd en faktor som belyses av flertalet av de intervjuade och undersökta personerna.

Nyckelord: cerebral pares, vuxna, funktion, hälsa, erfarenheter, självbild, autonomi, fysisk aktivitet, sjukgymnastik, uppföljning

## ACKNOWLEDGEMENTS

I wish to express my gratitude to a number of people who have encouraged and contributed to this thesis. In particular I want to thank all people who have attended long investigations and shared their experiences of living with cerebral palsy.

I also wish to express my sincere gratitude to the following people:

Birgitta Öberg, my supervisor, who has supported, encouraged and provided constructive criticism through the whole process. I admire your ability of analysis and comprehensive view.

Kersti Samuelsson, my co-supervisor. Your knowledge of the research area has inspired many discussions and your quick and clear answers have helped me in many situations.

Johan Alinder, my colleague at the Department of Rehabilitation Medicine. You started this process by giving me the opportunity to work with the first investigation. I had no idea what this could lead to.

My colleagues at the Department of Physiotherapy, for discussions, new ideas, practical help and encouragement! I particularly want to thank Pia Ödman for your support both in ups and downs. I also want to thank Emma Winquist for your help with layout of the final thesis.

Fredrik Svenaeus, for support in the initial stage of the interview study, and for your help in finding relevant research questions.

My colleagues at the Department of Rehabilitation Medicine at the University Hospital, for having the opportunity to work together with you all this years and for all enjoyments we have had together.

Benita Rennes, my former colleague in Child Habilitation, for drawing a cover illustration.

My family, for your constant support! From my parents I have learned what living with a disability means, but also that discussions and togetherness can override practical problems in daily life.

Financial support for this study was gratefully received from the County Council of Östergötland, Norrbacka-Eugenia Foundation, The Swedish Association of Persons with Neurological Disabilities, The Swedish Association of Registered Physiotherapists/Section of Neurology.

---

## REFERENCES

1. Mutch L, Alberman E, Hagberg B, Kodama K, Perat MV. Cerebral palsy epidemiology: Where are we now and where are we going? *Dev Med Child Neurol.* 1992; 34:547-551
2. Bax M, Goldstein M, Rosenbaum P, Leviton A, Paneth N, Dan B, Jacobsson B, Damiano D. Proposed definition and classification of cerebral palsy, april 2005. *Dev Med Child Neurol.* 2005; 47:571-576
3. Surveillance of Cerebral Palsy in Europe (SCPE). Prevalence and characteristics of children with cerebral palsy in Europe. *Dev Med Child Neurol.* 2002; 44:633-640
4. Beckung E, Hagberg G. Neuroimpairments, activity limitations, and participation restrictions in children with cerebral palsy. *Dev Med Child Neurol.* 2002; 44:309-316
5. Hagberg B, Hagberg G, Beckung E, Uvebrant P. Changing panorama of cerebral palsy in Sweden. VIII. Prevalence and origin in the birth year period 1991-94. *Acta Paediatr.* 2001; 90:271-277
6. Palisano R, Rosenbaum P, Walter S, Russell D, Wood E, Galuppi B. Development and reliability of a system to classify gross motor function in children with cerebral palsy. *Dev Med Child Neurol.* 1997; 39:214-223
7. Jahnsen R, Aamodt G, Rosenbaum P. Gross motor function classification system used in adults with cerebral palsy: Agreement of self-reported versus professional rating. *Dev Med Child Neurol.* 2006; 48:734-738
8. Palisano RJ, Rosenbaum P, Bartlett D, Livingston MH. Content validity of the expanded and revised gross motor function classification system. *Dev Med Child Neurol.* 2008; 50:744-750
9. Nordmark E, Hagglund G, Lagergren J. Cerebral palsy in southern Sweden I. Prevalence and clinical features. *Acta Paediatr.* 2001; 90:1271-1276
10. Himmelmann K, Hagberg G, Beckung E, Hagberg B, Uvebrant P. The changing panorama of cerebral palsy in Sweden. IX. Prevalence and origin in the birth-year period 1995-1998. *Acta Paediatr.* 2005; 94:287-294
11. Westbom L, Hägglund G, Nordmark E. Cerebral palsy in a total population of 4-11 year olds in southern Sweden. Prevalence and distribution according to different CP classification systems. *BMC Pediatr.* 2007; 7:41

12. Hemming K, Hutton JL, Pharoah PO. Long-term survival for a cohort of adults with cerebral palsy. *Dev Med Child Neurol.* 2006; 48:90-95
13. Blair E, Watson L, Badawi N, Stanley FJ. Life expectancy among people with cerebral palsy in western Australia. *Dev Med Child Neurol.* 2001; 43:508-515
14. Hutton JL, Pharoah PO. Life expectancy in severe cerebral palsy. *Arch Dis Child.* 2006; 91:254-258
15. Hutton JL, Colver AF, Mackie PC. Effect of severity of disability on survival in north east England cerebral palsy cohort. *Arch Dis Child.* 2000; 83:468-474
16. Strauss D, Shavelle R, Reynolds R, Rosenbloom L, Day S. Survival in cerebral palsy in the last 20 years: Signs of improvement? *Dev Med Child Neurol.* 2007; 49:86-92
17. Strauss D, Ojdana K, Shavelle R, Rosenbloom L. Decline in function and life expectancy of older persons with cerebral palsy. *NeuroRehabilitation.* 2004; 19:69-78
18. King G, Baldwin P, Currie M, Evans J. Planning successful transitions from school to adult roles for youth with disabilities. *Child Health Care.* 2005; 34:195-216
19. Rosenbaum P, Stewart D. Perspectives on transitions: Rethinking services for children and youth with developmental disabilities. *Arch Phys Med Rehabil.* 2007; 88:1080-1082
20. Stevenson CJ, Pharoah PO, Stevenson R. Cerebral palsy--the transition from youth to adulthood. *Dev Med Child Neurol.* 1997; 39:336-342
21. Fiorentino L, Datta D, Gentle S, Hall DM, Harpin V, Phillips D, Walker A. Transition from school to adult life for physically disabled young people. *Arch Dis Child.* 1998; 79:306-311
22. Stewart DA, Law MC, Rosenbaum P, Willms DG. A qualitative study of the transition to adulthood for youth with physical disabilities. *Phys Occup Ther Pediatr.* 2001; 21:3-21
23. Binks JA, Barden WS, Burke TA, Young NL. What do we really know about the transition to adult-centered health care? A focus on cerebral palsy and spina bifida. *Arch Phys Med Rehabil.* 2007; 88:1064-1073
24. Bachrach S, Greenspun B. Care of the adult with cerebral palsy. *Del Med J.* 1990; 62:1287-1290, 1293-1285
25. Murphy KP, Molnar GE, Lankasky K. Medical and functional status of adults with cerebral palsy. *Dev Med Child Neurol.* 1995; 37:1075-1084
26. Granet KM, Balaghi M, Jaeger J. Adults with cerebral palsy. *New Jersey Med.* 1997; 94:51-54

27. Turk MA, Geremski CA, Rosenbaum PF, Weber RJ. The health status of women with cerebral palsy. *Arch Phys Med Rehabil.* 1997; 78:S10-17
28. Murphy KP. Medical problems in adults with cerebral palsy: Case examples. *Assist Technol.* 1999; 11:97-104
29. Schwartz L, Engel JM, Jensen MP. Pain in persons with cerebral palsy. *Arch Phys Med Rehabil.* 1999; 80:1243-1246
30. Jahnsen R, Villien L, Stanghelle JK, Holm I. Fatigue in adults with cerebral palsy in Norway compared with the general population. *Dev Med Child Neurol.* 2003; 45:296-303
31. Jahnsen R, Villien L, Aamodt G, Stanghelle JK, Holm I. Musculoskeletal pain in adults with cerebral palsy compared with the general population. *J Rehabil Med.* 2004; 36:78-84
32. Hilberink SR, Roebroek ME, Nieuwstraten W, Jalink L, Verheijden JMA, Stam HJ. Health issues in young adults with cerebral palsy: Towards a life-span perspective. *J Rehabil Med.* 2007; 39:605-611
33. Maruishi M, Mano Y, Sasaki T, Shinmyo N, Sato H, Ogawa T. Cerebral palsy in adults: Independent effects of muscle strength and muscle tone. *Arch Phys Med Rehabil.* 2001; 82:637-641
34. Ando N, Ueda S. Functional deterioration in adults with cerebral palsy. *Clin Rehabil.* 2000; 14:300-306
35. Bottos M, Feliciangeli A, Sciuto L, Gericke C, Vianello A. Functional status of adults with cerebral palsy and implications for treatment of children. *Dev Med Child Neurol.* 2001; 43:516-528
36. Andersson C, Mattsson E. Adults with cerebral palsy: A survey describing problems, needs, and resources, with special emphasis on locomotion. *Dev Med Child Neurol.* 2001; 43:76-82
37. Jahnsen R, Villien L, Egeland T, Stanghelle JK, Holm I. Locomotion skills in adults with cerebral palsy. *Clin Rehabil.* 2004; 18:309-316
38. Andren E, Grimby G. Dependence in daily activities and life satisfaction in adult subjects with cerebral palsy or spina bifida: A follow-up study. *Disabil and Rehabil.* 2004; 26:528-536
39. van der Dussen L, Nieuwstraten W, Roebroek M, Stam HJ. Functional level of young adults with cerebral palsy. *Clin Rehabil.* 2001; 15:84-91
40. Michelsen SI, Uldall P, Kejs AM, Madsen M. Education and employment prospects in cerebral palsy. *Dev Med Child Neurol.* 2005; 47:511-517
41. Michelsen SI, Uldall P, Hansen T, Madsen M. Social integration of adults with cerebral palsy. *Dev Med Child Neurol.* 2006; 48:643-649

42. Schenker R, Coster W, Parush S. Participation and activity performance of students with cerebral palsy within the school environment. *Disabil Rehabil.* 2005; 27:539-552
43. Donkervoort M, Roebroek M, Wiegerink D, van der Heijden-Maessen H, Stam H. Determinants of functioning of adolescents and young adults with cerebral palsy. *Disabil Rehabil.* 2007; 29:453-463
44. Wiegerink DJ, Roebroek ME, Donkervoort M, Cohen-Kettenis PT, Stam HJ. Social, intimate and sexual relationships of adolescents with cerebral palsy compared with able-bodied age-mates. *J Rehabil Med.* 2008; 40:112-118
45. van der Slot WM, Roebroek ME, Landkroon AP, Terburg M, Berg-Emons RJ, Stam HJ. Everyday physical activity and community participation of adults with hemiplegic cerebral palsy. *Disabil Rehabil.* 2007; 29:179-189
46. van Meeteren J, Roebroek ME, Celen E, Donkervoort M, Stam HJ. Functional activities of the upper extremity of young adults with cerebral palsy: A limiting factor for participation? *Disabil Rehabil.* 2008; 30:387-395
47. Bent N, Jones A, Molloy I, Chamberlain MA, Tennant A. Factors determining participation in young adults with a physical disability: A pilot study. *Clin Rehabil.* 2001; 15:552-561
48. Rosenbaum PL, Livingston MH, Palisano RJ, Galuppi BE, Russell DJ. Quality of life and health-related quality of life of adolescents with cerebral palsy. *Dev Med Child Neurol.* 2007; 49:516-521
49. Livingston MH, Rosenbaum PL, Russell DJ, Palisano RJ. Quality of life among adolescents with cerebral palsy: What does the literature tell us? *Dev Med Child Neurol.* 2007; 49:225-231
50. Fresher-Samways K, Roush SE, Choi K, Desrosiers Y, Steel G. Perceived quality of life of adults with developmental and other significant disabilities. *Disabil Rehabil.* 2003; 25:1097-1105
51. Furukawa A, Iwatsuki H, Nshiyama M, Nii E, Uchida A. A study on the subjective well-being of adult patients with cerebral palsy. *J Phys Ther Sci.* 2001;13:31-35
52. Husserl E. *Fenomenologins idé* (in Swedish) Göteborg: Daidalos; 1989.
53. Svenaeus F. The phenomenology of health and illness. In: Toombs K, ed. *Handbook of phenomenology and medicine.* Dordrecht: Kluwer academic publishers; 2001.
54. Merleau-Ponty M. *Phenomenology of perception.* London: Routledge; 1962.



55. Castle K, Imms C, Howie L. Being in pain: A phenomenological study of young people with cerebral palsy. *Dev Med Child Neurol.* 2007; 49:445-449
56. King GA, Cathers T, Polgar JM, MacKinnon E, Havens L. Success in life for older adolescents with cerebral palsy. *Qual Health Res.* 2000; 10:734-749
57. Physical Activity Guidelines Advisory Committee. *Physical activity guidelines advisory committee report,2008.* Washington: Department of Health and Human Services; 2008.
58. World confederation of Physical Therapy (WCPT). *Position statement description of physical therapy.* London: 1999. <http://www.wcpt.org/publications/docs.php>
59. Yrkesföreningar för fysisk aktivitet (YFA). *Fysisk aktivitet i sjukdomsprevention och sjukdomsbehandling (FYSS).* Stockholm: Statens folkhälsoinstitut; 2008.
60. Andersson C, Grooten W, Hellsten M, Kaping K, Mattsson E. Adults with cerebral palsy: Walking ability after progressive strength training. *Dev Med Child Neurol.* 2003; 45:220-228
61. Taylor NF, Dodd KJ, Larkin H. Adults with cerebral palsy benefit from participating in a strength training programme at a community gymnasium. *Disabil Rehabil.* 2004; 26:1128-1134
62. Ahlborg L, Andersson C, Julin P. Whole-body vibration training compared with resistance training: Effect on spasticity, muscle strength and motor performance in adults with cerebral palsy. *J Rehabil Med.* 2006; 38:302-308
63. Allen J, Dodd KJ, Taylor NF, McBurney H, Larkin H. Strength training can be enjoyable and beneficial for adults with cerebral palsy. *Disabil Rehabil.* 2004; 26:1121-1127
64. Pitetti K, Fernandez J, Lanciault M. Feasibility of an exercise program for adults with cerebral palsy: A pilot study. *Adapt Phys Act Quart.* 1991; 8:333-341
65. Dodd KJ, Taylor NF, Damiano DL. A systematic review of the effectiveness of strength-training programs for people with cerebral palsy. *Arch Phys Med Rehabil.* 2002; 83:1157-1164
66. Ödman P, Öberg B. Effectiveness of intensive training for children with cerebral palsy--a comparison between child and youth rehabilitation and conductive education. *J Rehabil Med.* 2005; 37:263-270
67. Rimmer JH. Physical fitness levels of persons with cerebral palsy. *Dev Med Child Neurol.* 2001; 43:208-212

68. Liptak GS. Health and well being of adults with cerebral palsy. *Curr Opin Neurol.* 2008; 21:136-142
69. Jahnsen R, Villien L, Aamodt G, Stanghelle JK, Holm I. Physiotherapy and physical activity - experiences of adults with cerebral palsy, with implications for children. *Adv Physiother.* 2003; 5:21-32
70. Gaskin CJ, Morris T. Physical activity, health-related quality of life, and psychosocial functioning of adults with cerebral palsy. *J Physl Act& Health.* 2008; 5:146-157
71. Beatty PW, Hagglund KJ, Neri MT, Dhont KR, Clark MJ, Hilton SA. Access to health care services among people with chronic or disabling conditions: Patterns and predictors. *Arch Phys Med Rehabil.* 2003; 84:1417-1425
72. Heller T, Ying G, Rimmer JH, Marks BA. Determinants of exercise in adults with cerebral palsy. *Publ Health Nurs.* 2002; 19:223-231
73. Grimby G, Gudjonsson G, Rodhe M, Sunnerhagen KS, Sundh V, Ostensson ML. The functional independence measure in Sweden: Experience for outcome measurement in rehabilitation medicine. *Scand J Rehabil Med.* 1996; 28:51-62
74. Grimby G, Andren E, Holmgren E, Wright B, Linacre JM, Sundh V. Structure of a combination of functional independence measure and instrumental activity measure items in community-living persons: A study of individuals with cerebral palsy and spina bifida. *Arch Phys Med Rehabil.* 1996; 77:1109-1114
75. Grimby G, Andren E, Daving Y, Wright B. Dependence and perceived difficulty in daily activities in community-living stroke survivors 2 years after stroke: A study of instrumental structures. *Stroke.* 1998; 29:1843-1849
76. Hasselgren-Nyberg L, Omgren M, Nyberg L, Gustafson Y. Den svenska versionen av physiotherapy clinical outcome variables (in Swedish). *Nordisk Fysioterapi.* 1997; 1:109-113
77. Lindmark B, Hamrin E. Evaluation of functional capacity after stroke as a basis for active intervention. *Scand J Rehabil Med.* 1998; 20:103-115
78. Scott J, Huskisson EC. Graphic representation of pain. *Pain.* 1976; 2:175-184
79. Brooks R. Euroqol: The current state of play. *Health Policy.* 1996; 37:53-72
80. Kvale S. *Interviews : An introduction to qualitative research interviewing.* Thousand Oaks: SAGE; 1996.

81. Patton M. *Qualitative evaluations methods*. Newbury Park: SAGE Publications Inc; 2002.
82. Landis JR, Koch GG. The measurement of observer agreement for categorical data. *Biometrics*. 1977; 33:159-174
83. Karlsson G. *Psychological qualitative research from a phenomenological perspective*. Göteborg: Almqvist & Wiksell international; 1995.
84. Downe-Wamboldt B. Content analysis: Method, applications, and issues. *Health Care Women Int*. 1992; 13:313-321
85. Graneheim UH, Lundman B. Qualitative content analysis in nursing research: Concepts, procedures and measures to achieve trustworthiness. *Nurse Educ Today*. 2004; 24:105-112
86. Hsieh HF, Shannon SE. Three approaches to qualitative content analysis. *Qual Health Res*. 2005; 15:1277-1288
87. Medicinska forskningsrådet (MFR). *Riktlinjer för etisk värdering av medicinsk humanforskning*. Stockholm: Medicinska forskningsrådet; 1996.
88. Wood E, Rosenbaum P. The gross motor function classification system for cerebral palsy: A study of reliability and stability over time. *Dev Med Child Neurol*. 2000; 42:292-296
89. Nordmark E, Hägglund G, Lagergren J. Cerebral palsy in southern Sweden II. Gross motor function and disabilities. *Acta Paediatr*. 2001; 90:1277-1282
90. Himmelmann K, Beckung E, Hagberg G, Uvebrant P. Gross and fine motor function and accompanying impairments in cerebral palsy. *Dev Med Child Neurol*. 2006; 48:417-423
91. Lincoln Y, Guba E. Establishing trustworthiness. In: Bryman A, Burgess R, eds. *Qualitative research, vol III*. Sage publications; 1999.
92. Höijer B. Reliability, validity and generalizability. *Nordicom Rev*. 1990; (1): 15-20
93. Hammersley M. *What's wrong with ethnography?* London: Routledge; 1995.
94. McCormick A, Brien M, Plourde J, Wood E, Rosenbaum P, McLean J. Stability of the gross motor function classification system in adults with cerebral palsy. *Dev Med Child Neurol*. 2007; 49:265-269
95. Rosenbaum PL, Palisano RJ, Bartlett DJ, Galuppi BE, Russell DJ. Development of the gross motor function classification system for cerebral palsy. *Dev Med Child Neurol*. 2008; 50:249-253
96. Leder D. *The absent body*. Chicago: The university of Chicago press; 1990.

97. Engel JM, Jensen MP, Hoffman AJ, Kartin D. Pain in persons with cerebral palsy: Extension and cross validation. *Arch Phys Med Rehabil.* 2003; 84:1125-1128
98. Engel JM, Kartin D, Jensen MP. Pain treatment in persons with cerebral palsy: Frequency and helpfulness. *Am J Phys Med Rehabil.* 2002; 81:291-296
99. Jensen MP, Engel JM, Schwartz L. Coping with cerebral palsy pain: A preliminary longitudinal study. *Pain Med.* 2006; 7:30-37
100. Jahnsen R, Villien L, Stanghelle JK, Holm I. Coping potential and disability--sense of coherence in adults with cerebral palsy. *Disabil Rehabil.* 2002; 24:511-518
101. Andren E, Grimby G. Dependence in daily activities and life satisfaction in adult subjects with cerebral palsy or spina bifida: A follow-up study. *Disabil Rehabil.* 2004; 26:528-536
102. Furukawa A. A study on the subjective well-being of adult patients with cerebral palsy. *J Phys Ther Sci.* 2001; 13:31-35
103. Albrecht GL, Devlieger PJ. The disability paradox: High quality of life against all odds. *Soc Sci Med.* 1999; 48:977-988
104. Eriksson E, Nordlund A. *Hälsa och hälsorelaterad livskvalitet mätt med EQ-5D och SF-36 i Östergötlands och Kalmar län: Resultat från befolkningsenkäterna* (in Swedish). Linköping: Folkhälsovetenskapligt centrum; 2002.
105. Turk MA, Scandale J, Rosenbaum PF, Weber RJ. The health of women with cerebral palsy. *Phys Med Rehabil Clin N Am.* 2001; 12:153-168
106. Livingston MH, Rosenbaum PL. Adolescents with cerebral palsy: Stability in measurement of quality of life and health-related quality of life over 1 year. *Dev Med Child Neurol.* 2008; 50:696-701
107. Bury M. Chronic illness as biographical disruption. *Sociol Health Illn.* 1982; 4:167-182
108. Toombs S. *The meaning of illness.* Dordrecht: Kluwer academic press; 1992.
109. Wendell S. *The rejected body.* New York: Routledge; 1996.
110. Goffman E. *Jaget och maskerna* (Original title: the presentation of self in everyday life, 1959). Stockholm: Norstedts akademiska förlag; 2004.
111. Taleporos G, McCabe MP. Body image and physical disability--personal perspectives. *Soc Sci Med.* 2002; 54:971-980
112. Samonds RJ, Cammermeyer M. Perceptions of body image in subjects with multiple sclerosis: A pilot study. *J Neurosci Nurs.* 1989; 21:190-194

113. Lo Surdo P. Cerebral palsy and ageing. A personal account. *Aust Fam Physician*. 1997; 26:404-407
114. Barron K. *The transition from adolescence to adulthood for physically disabled young people*. Uppsala: Centre for disability research, Uppsala university; 1995.
115. Adamson L. Self-image, adolescence, and disability. *Am J Occup Ther*. 2003; 57:578-581
116. Antle BJ. Factors associated with self-worth in young people with physical disabilities. *Health Soc Work*. 2004; 29:167-175
117. Jonsson G, Ekholm J, Schult ML. The international classification of functioning, disability and health environmental factors as facilitators or barriers used in describing personal and social networks: A pilot study of adults with cerebral palsy. *Int J Rehabil Res*. 2008; 31:119-129
118. Becker H, Schaller J. Perceived health and self-efficacy among adults with cerebral palsy. *J Rehabil*. 1995; 61:36-42
119. Bandura A. *Self-efficacy. The exercise of control*. New York: WH Freeman & Co; 1997.
120. MacKenzie C. On bodily autonomy. In: Toombs, K ed. *Handbook of phenomenology and medicine*. Dordrecht: Kluwer academic publishers; 2001.
121. Kibele A. Occupational therapy's role in improving the quality of life for persons with cerebral palsy. *Am J Occup Ther*. 1989; 43:371-377
122. Ekenberg L. *Att vara barn med rörelsehinder- upplevelser av sjukgymnastik. Intervjuer med unga vuxna i Norrbotten* (in Swedish). Luleå: FoU-rapport. 2004
123. Fernandez JE, Pitetti KH, Betzen MT. Physiological capacities of individuals with cerebral palsy. *Hum Factors*. 1990; 32:457-466
124. Goodwin D, Compton S. Physical activity experiences of women aging with disabilities. *Adapt Phys Act Quart*. 2004; 21:122-138
125. Zabriskie R, Lundberg N, Groff D. Quality of life and identity: The benefits of a community-based therapeutic recreation and adaptive sports program. *Ther Recr J*. 2005; 39:176-191
126. Ellis R, Kosma M, Cardinal BJ, Bauer JJ, McCubbin JA. Physical activity beliefs and behaviour of adults with physical disabilities. *Disabil Rehabil*. 2007; 29:1221-1227
127. Yuen HK, Hanson C. Body image and exercise in people with and without acquired mobility disability. *Disabil Rehabil*. 2002; 24:289-296
128. Palisano RJ, Copeland WP, Galuppi BE. Performance of physical activities by adolescents with cerebral palsy. *Phys Ther*. 2007; 87:77-87

129. Rimmer JH. Health promotion for people with disabilities: The emerging paradigm shift from disability prevention to prevention of secondary conditions. *Phys Ther.* 1999; 79:495-502
130. Ng SY, Dinesh SK, Tay SK, Lee EH. Decreased access to health care and social isolation among young adults with cerebral palsy after leaving school. *J Orthop Surg (Hong Kong).* 2003; 11:80-89
131. Neri MT, Kroll T. Understanding the consequences of access barriers to health care: Experiences of adults with disabilities. *Disabil Rehabil.* 2003; 25:85-96
132. Young NL, Steele C, Fehlings D, Jutai J, Olmsted N, Williams JI. Use of health care among adults with chronic and complex physical disabilities of childhood. *Disabil Rehabil.* 2005; 27:1455-1460
133. Young NL, Gilbert TK, McCormick A, Ayling-Campos A, Boydell K, Law M, Fehlings DL, Mukherjee S, Wedge JH, Williams JI. Youth and young adults with cerebral palsy: Their use of physician and hospital services. *Arch Phys Med Rehabil.* 2007; 88:696-702
134. Van Naarden Braun K, Yeargin-Allsopp M, Lollar D. A multi-dimensional approach to the transition of children with developmental disabilities into young adulthood: The acquisition of adult social roles. *Disabil Rehabil.* 2006; 28:915-928
135. Magill-Evans J, Wiart L, Darrah J, Kratochvil M. Beginning the transition to adulthood: The experiences of six families with youths with cerebral palsy. *Phys Occup Ther Pediatr.* 2005; 25:19-36
136. Svien LR, Berg P, Stephenson C. Issues in aging with cerebral palsy. *Topics Ger Rehabil.* 2008; 24:26-40
137. The Roeher Institute. *Aging and CP.* Ontario federation for cerebral palsy; 1996. <http://www.ofcp.on.ca/aging.html>
138. King G, Willoughby C, Specht JA, Brown E. Social support processes and the adaptation of individuals with chronic disabilities. *Qual Health Res.* 2006; 16:902-925
139. Oliver M. *Understanding disability.* New York: St Martin's press; 1996.
140. Rejeski WJ, Focht BC. Aging and physical disability: On integrating group and individual counselling with the promotion of physical activity. *Exerc Sport Sci Rev.* 2002; 30:166-170
141. Pueyo R, Junque C, Vendrell P. Neuropsychologic differences between bilateral dyskinetic and spastic cerebral palsy. *J Child Neurol.* 2003; 18:845-850

- 
142. Sigurdardottir S, Eiriksdottir A, Gunnarsdottir E, Meintema M, Arnadottir U, Vik T. Cognitive profile in young Icelandic children with cerebral palsy. *Dev Med Child Neurol.* 2008; 50:357-362
  143. Cooper RA, Quatrano LA, Axelson PW, Harlan W, Stineman M, Franklin B, Krause JS, Bach J, Chambers H, Chao EY, Alexander M, Painter P. Research on physical activity and health among people with disabilities: A consensus statement. *J Rehabil Res Dev.* 1999; 36:142-154
  144. Rimmer JH, Braddock D, Pitetti KH. Research on physical activity and disability: An emerging national priority. *Med Sci Sports Exerc.* 1996; 28:1366-1372