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Abstract

In children with cerebral palsy (CP) spastic diplegia, spasticity is a common motor impairment affecting movement quality and may lead to secondary musculoskeletal impairments such as muscle contractures and joint deformities. Selective Dorsal Rhizotomy (SDR) is a permanent spasticity reducing operation which in combination with physiotherapy treatment has been shown to improve functional outcomes in children with spastic diplegia.

The aim of this thesis was to describe the development of children's functional outcomes over the course of 10 years following SDR combined with physiotherapy and to evaluate measures used in follow-up.

This thesis comprises five original papers. Functional outcomes from children with spastic diplegia undergoing SDR at the University Hospital in Lund, Sweden, between 1993- 2004 were included in this work.

Functional outcomes have been monitored by standardized measures in accordance to the components of International Classification of Functioning Disability and Health-Children and Youth (ICF-CY) body functions and body structures: muscle tone (Papers I and V), passive range of motion (Papers I and V), reflexes and clonus (Paper I) and three- dimensional gait analysis (3DGA)(Paper IV). In the ICF components of activity and participation the following measures were used: capacity of gross motor function with Gross Motor Function Measure (GMFM-88 total and goal total scores (Papers I, III) and GMFM-66 (Papers I, III-V)), performance of functional skills and caregivers' assistance in self care and mobility with scaled scores of the Pediatric Evaluation Disability Inventory (PEDI) (Paper I) and functional mobility with the Functional Mobility Scale (FMS) (Paper V). The gross motor function of all children was classified according the Gross Motor Function Classification System (GMFCS) in a five level scale according to the severity of functional limitations.

In Papers I and II functional outcomes in 35 children with a mean age at SDR of 4.5 years and who were followed for five years were included. In Paper V, functional outcomes in 29 adolescents with a mean age at the 10-year follow-up of 14.8 years were studied. Muscle tone was immediately reduced and the reduction was maintained at five and 10 years after SDR. The passive range of motion improved during the first five years after SDR. However, at 10 years after SDR the mean PROM was only slightly different to the preoperative, indicating a decrease between 5 and 10 years postoperatively. Less than half of the children had undergone orthopaedic surgery after five years whilst stabilizing procedures of the foot and lengthening of achilles and hip adductor muscle tendons were the most common

interventions. Children with walking capacity most often had surgery of the foot and children without walking capacity had mostly surgical interventions of the hip.

The improvement in gross motor function was seen at 12 months after SDR and was then continued during five years after SDR. At 10 years after SDR, changes were still improved compared with preoperative status. Children with less functional limitations and children operated on at younger ages experienced larger changes in capacity of gross motor function compared to children with larger functional limitations and children operated on at older ages. After five years, all children performed more functional skills and were more independent in daily activities of self care and mobility than before SDR measured with scaled scores of the PEDI. At 10 years after SDR performance of functional mobility among the adolescents varied according to functional limitations and contexts according to the FMS.

In Paper III, GMFM scores were included from 41 children undergoing SDR at a mean age of 4.4 years and followed for five years. The three GMFM versions, namely GMFM-88 total, goal total and GMFM-66 scores, were examined for longitudinal construct validity in Paper III. GMFM-88 total and goal score indicated large changes earlier than GMFM-66, whilst large changes were seen in all scoring options over the five years. GMFM-66 identified changes in children with less motor impairment as well as in children with larger functional limitations in spite of the reduced number of items in lying, sitting and kneeling positions.

In Paper IV, 3DGA was performed in seven children before and at one, three and five years after SDR. Selected 3DGA parameters were used to obtain Gait Profile Scores (GPS) and Movement Assessment Profiles (MAP). The GPS showed an improved overall gait score in six of the seven children and unchanged in one child during the five years. The MAP illustrated decreased deviation of the angles of the dorsiflexion and foot progression in one selected child during the five years.

In this thesis, SDR combined with physiotherapy have been shown to provide immediate, safe and effective spasticity reduction after 10 years. The mean passive range of motion was increased during the first five years and between five and 10 years after SDR a decrease was seen. Improvement in mean gross motor function for the whole group was seen after five and 10 years. The children performed more functional skills and were more independent in self care and mobility at five years after SDR compared to preoperative scores. Changes in GMFM-66 scores between pre- and 10 years postoperatively depended on preoperative age and the severity of functional limitations. Performance of functional mobility was related to the severity of CP and contexts at 10 years after SDR.

All three scoring options available from the GMFM identified large changes and postoperatively the GMFM-88 total and goal total scores detected large changes earlier postoperatively. GMFM-66 could identify changes in gross motor function in

children in GMFCS levels I-III and IV-V. The GPS and MAP may become a helpful tool in interpreting and communicating 3DGA results. The GPS and MAP were found useful for longitudinal studies with repeated sessions.

The intervention SDR, combined with physiotherapy, has been shown to affect functional outcomes in both the components of body functions and body structures as well as in activity and participation. A combination of standardized measures covering these aspects should be used to identify important changes in functional outcomes.

Summary in Swedish

Svensk sammanfattning

Hos barn med cerebral pares (CP) spastisk diplegi är förhöjd muskelspänning s.k. spasticitet är ett vanligt symptom som påverkar kvalitén på rörelser. Muskler blir stela och förkortas och felställningar i leder kan uppstå. Selektiv Dorsal Rhizotomi (SDR) är ett ingrepp som används för att minska spasticitet i benen och som i kombination med sjukgymnastik kan ge bestående funktions förbättringar.

Syftet med denna avhandling var att beskriva långtidsresultat hos barn med CP som genomgått SDR i kombination med sjukgymnastik samt att undersöka mätmetoder som kan användas vid uppföljningen för dessa barn.

Avhandlingen omfattar fem delarbeten. I dessa arbeten ingår resultat från mätningar av funktionsförmåga från barn med spastisk diplegi som genomgick SDR i Lund mellan åren 1993 och 2004.

Barnens funktionsförmåga har följts upp med standardiserade mätinstrument för de båda komponenterna kroppsfunktion och kroppsstruktur samt aktivitet och delaktighet enligt International Classification of Functioning Disability and Health-Children and Youth version (ICF-CY). Bedömning av funktionsförmåga i komponenten kroppsfunktioner och kroppsstrukturer gjordes med följande mätmetoder; skattning av muskelspänning (delarbetena I, V), reflexer (delarbete I), ledrörlighet i passivt rörelseuttag (delarbetena I, V) samt tredimensionell gånganalys (delarbete IV). Bedömning av funktionsförmåga i komponenten aktiviteter och delaktighet gjordes med följande mätmetoder; grovmotorisk funktionsförmåga med Gross Motor Function Measure (GMFM-88 total, goal total (delarbetena I, III) och GMFM-66(delarbetena I, III-V)), utförande av funktionella färdigheter och hjälpbehov vid personlig vård och rörelseförmåga med skalpoäng för Pediatric Evaluation Disability Inventory (PEDI) (delarbete I) samt utförande av funktionell förflyttning med Functional Mobility Scale (FMS) (delarbete V). Den preoperativa grovmotoriska funktionsförmågan klassificerades med Gross Motor Function Classification System (GMFCS), en femgradig skala där nivå I representerar liten och nivå V representerat en stor funktions begränsning (delarbetena I-V).

I delarbetena I och II presenteras resultat fem år efter SDR från 35 barn med en medelålder vid SDR av 4.5 år. I delarbete V presenteras 10 års resultat från 29 ungdomar som opererades vid en medelålder av 4.5 år och de var 14.8 år vid återbesöket 10 år efter SDR. Spasticiteten minskade direkt efter operationen och var fortsatt reducerad efter fem och 10 år. I genomsnitt förbättrades ledrörligheten i höft, knä och fot de första fem åren. Tio år efter SDR hade ledrörligheten för gruppen

minskat i höft, knä och fot till värden i stort sett motsvarande de från före operationen. Barn med större grad av funktionsbegränsningar försämrade ledrörlighet kring höft och knä mer än barn med mindre funktionsbegränsningar. Däremot förbättrades ledrörligheten i fotleden för barn med större grad av funktionsbegränsning i större utsträckning än för barnen med mindre funktionsbegränsningar.

Knappt hälften av alla barn hade genomgått någon typ av ortopedisk operation fem år efter SDR. De vanligaste ingreppen var stabilisering av foten samt förlängningar av muskelsenor kring fot- och höftled. Barn som kunde gå självständigt opererades till största delen i fötterna medan barn utan självständig gångförmåga opererades kring höftleden. Grovmotorisk funktion var förbättrad vid fem år. Tio år efter SDR fanns fortfarande säkerställda förbättringar i jämförelse med före operationen. Storleken på förändringarna mellan mätningarna före och 10 år efter SDR berodde på ålder och grad av funktionella begränsningar vid SDR. Yngre barn med mindre funktionsbegränsningar förbättrades mest med GMFM-66.

Fem år efter SDR utförde barnen fler funktionella färdigheter och var mer självständiga vid aktiviteter inom personlig vård och rörelseförmåga utifrån PEDI resultat. Tio år efter SDR sågs funktionell förflyttningsförmåga bero på svårighetsgrad av funktionsbegränsning samt omgivningsfaktorer utifrån FMS.

I delarbete III ingår resultat från 41 barn som genomgick SDR vid en medelålder av 4.4 år och som följts under fem år. De tre olika GMFM versionerna GMFM-88 total, goal total och GMFM-66 visade små förändringar efter 6 månader. Därefter successivt ökade förändringarna upp till fem år efter SDR. GMFM-88 total och goal total poängen identifierade stora förändringar tidigare i förloppet än GMFM-66. GMFM-66 kunde påvisa förändringar för både barn med stora såväl som för barn med mindre funktionella begränsningar, trots att instrumentet innehöll färre moment i liggande, sittande och knästående.

I delarbete IV ingår resultat från samtliga sju barn som följts under fem år med tredimensionell gånganalys. Medelålder vid SDR var 5.5 år. Utvalda resultat från den tredimensionella gånganalysen användes för att ta fram Gait Profile Score (GPS) och Movement Assessment Profile (MAP). GPS påvisade normaliserat gångmönster hos sex av sju samt oförändrat gångmönster hos ett barn. MAP visade på mindre avvikelse fotledsvinkeln och i fotens vinkel i förhållande till gångriktningen. Största förändringen i GPS och MAP sågs efter ett år.

Denna avhandling visar att SDR i kombination med sjukgymnastik ger en säker och effektiv minskning av spasticitet för barn med spastisk diplegi. Ledrörlighet förbättrades i genomsnitt för gruppen de första fem åren men 10 år efter operationen hade den minskat i höft och knä. Grovmotorisk funktion förbättrades under de första fem åren och vid 10 år fanns förbättringar jämfört med innan operationen. Fem år

efter SDR utförde barnen fler funktionella färdigheter och hade ett minskat hjälpbehov för moment inom personlig vård och rörelseförmåga mätt med PEDI. För grovmotorisk funktion kunde GMFM-88 identifiera stora förändringar vid ett tidigare skede efter SDR än GMFM-66. GMFM-66 kunde påvisa förändringar för både barn med stora såväl som för barn med mindre funktionella begränsningar. GPS och MAP ansågs vara användbara för att åskådliggöra utveckling av gångmönster vid uppföljning med flera mättillfällen hos barn som genomgått SDR.

SDR i kombination med sjukgymnastik har visats påverka funktionsförmågan inom ICF komponenterna kroppsstruktur, kroppsfunktion, aktivitet och delaktighet. Genom att kombinera olika standardiserade mätmetoder för de olika komponenterna vid uppföljning efter SDR kan förändringar i funktionsförmåga påvisas.

Thesis at a glance

Paper I: Long-term follow-up five years after SDR (n=35)	
Aim	To evaluate long-term functional outcomes, safety and side effects during 5 years after SDR.
Methods	Reflexes, clonus, muscle tone, PROM, GMFM-88, GMFM-66 and PEDI monitored changes.
Results	Muscle tone was immediately reduced, PROM was increased in hip, knee and foot, GMFM and PEDI increased.
Conclusion	SDR is safe and effective. Combined with physiotherapy treatment it provides lasting functional benefits during 5 years.
Paper II: Orthopaedic surgery after SDR (n=35)	
Aim	To analyze the amount and types of orthopaedic surgery 5 years after SDR.
Methods	Data was collected from medical reports at pre- and 5 years postoperatively.
Results	15 children underwent surgery. Subtalar arthodesis, achilles and adductor tendon lengthening were the most frequent operations.
Conclusion	Less than half of the children had had orthopaedic surgery. Stabilizing surgery of the foot and tenotomies of achilles and adductor muscle tendons were the most common operations.
Paper III: Longitudinal construct validity (LCV) of the GMFM-88 total, goal total and GMFM-66 scores (n=41)	
Aim	To study the LCV of the three GMFM scoring options.
Methods	ES and SRM were calculated for changes between pre- and 6, 12 and 18 months, as well as 3 and 5 years postoperatively.
Results	Large ES and SRM at 12 months postop were seen for GMFM-88 total and goal total scores for GMFM-66 scores at later follow-up.
Conclusion	All three options showed large LCV during follow-up, GMFM-66 could identify changes in gross motor function for children in GMFCS levels I-III and IV-V.
Paper IV: Gait profile Score (GPS) and Movement Assessment Profile (MAP) in a longitudinal follow-up (n=7)	
Aim	To explore the utility of GPS and MAP in a long-term follow-up in children with CP.
Methods	GPS and MAP were obtained from 3DGA data from pre-, 1, 3 and 5 years postoperatively.
Results	Changes in gait pattern over the 5 years were shown by GPS and MAP.
Conclusion	GPS and MAP were found to be useful for longitudinal studies with repeated sessions.
Paper V: Functional outcomes 10 years after SDR (n= 29)	
Aim	To describe changes in muscle tone, PROM and GMFM-66 between pre- and 10 years postop, to identify factors that could explain the changes and describe functional mobility at 10 years postop.
Methods	Muscle tone, PROM and GMFM-66 were analyzed according to GMFCS level and age, pre- to 10 years postop, FMS was used at 10 years postop.
Results	Muscle tone was continuously reduced. Mean PROM was similar to preop. GMFM-66 increased, changes were related to GMFCS levels and age. FMS identified variation in mobility at 10 years.
Conclusion	Muscle tone reduction was maintained, changes in mean PROM were small. Gross motor function improved, influenced by preop GMFCS levels and age. Functional mobility depended on GMFCS- E&R level and contexts.

Passive Range of Motion (PROM), postoperatively (postop), Gross Motor Function Measure (GMFM), Pediatric Evaluation Disability Inventory (PEDI), Effect Size (ES), Standardized Response Mean (SRM), Functional Mobility Scale (FMS). Gross Motor Function Classification System- Expanded & Revised (GMFCS- E&R)

Abbreviations

3DGA	Three Dimensional Gait Analysis
Btx	Botulinum Toxin A
CP	Cerebral Palsy
CPUP	Quality register and follow-up program of children with Cerebral Palsy
ES	Effect Size
EMG	Electromyography
FMS	Functional Mobility Scale
GMAE	Gross Motor Ability Estimator
GMFCS	Gross Motor Function Classification System
GMFCS-E&R	Gross Motor Function Classification System- Expanded and Revised
GMFM	Gross Motor Function Measure
GPS	Gait Profile Score
ICF	International Classification of Functioning, Disability and Health
ICF-CY	International Classification of Functioning, Disability and Health. Children & Youth Version
MAP	Movement Assessment Profile
OMG	Ontario Motor Growth
PEDI	Pediatric Evaluation of Disability Inventory
PROM	Passive Range of Motion
SDR	Selective Dorsal Rhizotomy
SRM	Standardized Response Mean
SVMC	Selective Voluntary Motor Control
WHO	World Health Organization

Definitions

Activity- The execution of a task or action by an individual [1].

Assessment- The systematic acquisition of information that is relevant and meaningful in providing the clinician with a comprehensive picture of the patient's abilities and problems [2].

Cerebral palsy- Definition by Mutch et al. "an umbrella term covering a group of non-progressive, but often changing, motor impairment syndromes secondary to lesions or anomalies of the brain arising in the early stages of its development."[3, p. 549].

Capacity- What the person can do in a standardized, controlled environment [4].

Capability- What the person can do in a daily environment [4].

Construct validity- The extent to which predefined hypothesized associations among different measures of the concept are confirmed [5].

Effect size (ES)- Mean difference between the baseline score and the follow-up scores divided by the standard deviation of the baseline score [6].

Function- The special, normal or proper physiological activity of an organ or part [7].

Impairment- Problems in body function or structure such as significant deviation or loss [1].

Inter-rater reliability- Consistency of measurements between raters [5].

Intra-rater reliability- Consistency of measurements between measurements recorded by the same rater [5].

Kinematics- Quantitative description of motion presented for clinical gait analysis as the time histories of angular displacement i.e. segment and joint angles over the gait cycle [8].

Kinetics- The effect of forces and torques on the motion of bodies [8].

Longitudinal construct validity (LCV)- The extent to which an instrument can detect a purposive change longitudinally within the construct it is intended to measure [9].

Measurement- The determination expressed numerically of the extent or quality of a substance, energy, or time [10].

Muscle strength- The capacity of a muscle to produce the tension necessary for maintaining posture, initiating movement, or controlling movement during conditions of loading on the musculoskeletal system[11].

Muscle tone- The force with which the muscle resists being lengthened, that is, stiffness [12].

Outcome- The condition of a client at the end of therapy or of a disease process, including the degree of wellness and the need for continuing care, medication, support, counseling or education [10]

Participation- The involvement in a life situation [1].

Performance- What a person actually does do in a daily life situation [4].

Range of motion- The range, measured in degrees of a circle, through which a joint can be extended and flexed [7].

Reliability- Reflect the amount of error, both random and systematic, inherent in any measurement [5].

Responsiveness- The ability of an instrument to measure a meaningful or clinically important change in a clinical state [9].

Sensitivity to change- The ability of an instrument to measure change in a state regardless of whether it is relevant or meaningful to the decision maker [9].

Spastic diplegic cerebral palsy- The subtype of CP with greater involvement of spasticity and impaired function in the legs than in the arms [13]. In the thesis the term "spastic diplegia" will be used.

Spasticity- "a motor disorder characterized by a velocity dependent increase in tonic stretch reflex with exaggerated tendon jerks, resulting from hyperexcitability of the stretch reflex, and is one component of the upper motor neuron syndrome"[14, p 498].

Standardized Response Mean (SRM)- Mean change score divided by the standard deviation of the change score [6].

Temporo-spatial parameters- Parameters pertaining to both time and space [7].

Type II error- A statistical error in which it is concluded that there is no difference between groups when, in fact, there is a difference [15].

Introduction

Cerebral Palsy (CP)

CP is the most common cause of motor disability in childhood in the western world, with an incidence of 2-3 /1000 births [16-19].

Over the last two centuries, different definitions of cerebral palsy have been discussed. The definition by Mutch et al. published in 1992, has frequently been used and will be the definition used in this thesis [3]. CP is defined as "an umbrella term covering a group of non-progressive, but often changing, motor impairment syndromes secondary to lesions or anomalies of the brain arising in the early stages of its development."[3, p. 549]. The central part is that the injury causing the brain dysfunction has occurred at an early age, before the age of two years, and that it is stationary while the movement restrictions are changing throughout life.

In Sweden the classification of CP by Hagberg et al. has been commonly used since the 1950-ies [13]. The three main types according to the dominating neurological symptoms are the spastic, dyskinetic and ataxic forms present in 83%, 12% and 4% respectively [19]. The spastic form can be subdivided into hemiplegia, diplegia and tetraplegia, depending on its distribution of spasticity. Spastic tetraplegia has equal or greater involvement in the arms than in the legs in contrast to compared to diplegia with greater involvement in the legs; hemiplegia involves one side of the body. The ataxic form is either diplegic or simple ataxia and the dyskinetic form is either mainly dystonic or mainly choreoathetotic. In a few children a single dominating symptom is impossible to decide, referred to as mixed subtype. The Hagberg classification will be used in this thesis.

The children included in this thesis have the CP subtype of spastic diplegia. It is the most common subtype together with spastic hemiplegia, each present in approximately one out of three of a total CP population using the Hagberg classification [16, 20]. However, spastic diplegia is a heterogeneous group with functional limitations throughout the whole spectrum, from minor to severe. Sixty percent of children with spastic diplegia walk without walking aids, 20% walk with walkers or crutches and the rest rely on wheelchairs for mobility [16].

Spastic diplegia is the typical CP subtype in children with periventricular leucomalacia, haemorrhage and/or infarction (PVL/PVH) with damage of the long descending axons of the upper motor neurons. This loss of white matter occurs most

often during the late second and early third trimester of the pregnancy (gestational weeks 24-34). The majority of children with PVL/PVH and spastic diplegia are born prematurely [21].

Upper motor neuron symptoms in children with spastic CP

Symptoms originating from injuries in the upper motor neuron are usually divided into positive symptoms characterized by a release of abnormal responses, or negative symptoms, characterized by the loss of normal responses. Positive symptoms are spasticity, increased flexor reflexes, secondary musculoskeletal symptoms and remaining developmental reactions. Decreased central dyscoordination, paresis and limited endurance are examples of negative symptoms [22].

Lance defined spasticity as "a motor disorder characterized by a velocity dependent increase in tonic stretch reflex with exaggerated tendon jerks, resulting from hyperexcitability of the stretch reflex, and is one component of the upper motor neuron syndrome" [14, p. 498].

Spasticity contributes to a major part of movement dysfunction in children with CP and has been shown to be present in 75-80 % of cases in a total population of children with CP [17, 23]. Spasticity occurs as a result of the decreased inhibition of the spinal flexor reflex. An increased excitability in the spinal extensor reflexes causes easily elicited and strong muscle tendon reflexes and clonus.

Spasticity will vary according to the time that has passed since the brain injury and the state of arousal. Initially a flaccid and inexcitable phase is seen, followed by a phase of hyperreflexia and clonus followed by reduced reflex excitability, and in the last phase muscles are stiff, inexciteable and shortened [24].

The negative symptoms are often underestimated; they are of large importance with respect to how motor function develops after the injury and contain central dyscoordination and paresis. Central dyscoordination means the inability to perform well coordinated movements where the joints moving independently of each other. Instead, stereotyped movements of the legs or arms are performed in a flexor or extensor synergistic movement pattern. As part of the deficient coordination, an increased co-activation of antagonist muscles occurs as a result of the lack of reciprocal inhibition. As the prerequisites of coordinated movements are both biomechanical and neuromuscular, peripheral factors, such as secondary neuromuscular impairments, may also contribute to the loss of coordinated functional movements [25].

Another important negative symptom is paresis or muscle weakness in the voluntary gross motor function. Movements are slow, cannot develop as much force as expected in peers without motor disability, and the muscles are more easily worn out. Paresis has been suggested to be most prominent distally in spastic diplegia and with an imbalance between agonist and antagonist [26, 27]. A relationship between increased muscle strength, improved temporo-spatial parameters, improved joint excursions during gait and capacity in gross motor function has been shown [28, 29]. During the last decade, increased strength, after different strength training programs, resulting in improved Gross Motor Function Measure (GMFM) scores, has been reported [30-32].

Secondary adaptive processes in the peripheral musculoskeletal systems occur as a result of the primary impairment in the central nervous system. These secondary conditions are often a focus for physiotherapy treatment.

Hypoextensibility in muscles with spasticity has been shown to depend on a reduced number of sarcomeres, impaired muscle growth and stiffening of elastic structures [33-35]. Changes in rectus femoris and vastus lateralis were shown to be different in children and adolescents with CP compared to typically developing peers. Changes were similar to those seen in disuse and ageing [36].

Also paresis will result in alteration of the muscle structure. If the muscle is positioned in a shortened position, it will be unloaded. Unloading of the muscle is the first step towards a contracture. The unloading of a muscle results in loss of muscle mass, seen as a loss of cross sectional muscle fiber area and sarcomeres, an increase of connective tissue and the accumulation of fat deposits in tendons. However, when the muscle is positioned in an extended position, less atrophy and an increase in the number of sarcomeres in series are seen [37].

Muscle contractures affect motor behaviour in children with CP, and joint contractures may also interfere with the acquisition of motor abilities. Nordmark et al. showed a decrease in PROM in popliteal angle, hip abduction, external rotation of the hip, knee extension and dorsiflexion of the ankle in children with spastic bilateral CP between the ages two and 14 years in total population [38]. The speed of development of contractures depends mostly on the severity of the motor disorder and the rate of physical growth [39].

Aerobic capacity, muscle strength, anaerobic muscle power and agility are often reduced in children with CP, which also affects their daily physical functioning [27, 40-42].

Limitations in motor function in cerebral palsy are often associated and co-exist with various other problems. Learning disability, epilepsy, difficulties in speech, hearing and vision are commonly associated impairments [43, 44]. In a total population of children with spastic diplegia, two out of three had cognitive functions within the

limits of typically developing children. Epilepsy was present in one out of four of the children and severe visual impairments in one out of five [23].

Gait in children with spastic diplegia

Before they can walk unsupported, children with spastic diplegia exhibit a gait pattern similar to that of typically developing children. However, as they mature, some of the characteristics of the infant stepping pattern, such as synchronous muscle activity with excessive muscular co-contraction and short-latency reflexes at foot contact have been found to be maintained [45].

The most common postures found in a large cohort with children with spastic diplegia were that of a stiff knee, flexed gait (also known as crouch gait), excess hip flexion, in toeing and equinus. Increased calcaneus and rotational malalignments were present in all subtypes of CP in higher ages and children with spastic diplegia exhibited commonly increased knee flexion in higher ages [46].

Longitudinal studies documenting changes in gait patterns show deterioration such as decreased excursions at the different joints of the lower limb, as well as temporo-spatial parameters of the gait [47-50].

Habilitation for children with CP

In Sweden, children diagnosed with CP are referred to local habilitation centers to receive specialized interventions. The habilitation centers provide multi-professional services for children, adolescents and adults with life-long functional limitations, with the aim to reach their best possible functional ability to gain full participation in the community. The work in the habilitation team is based on a comprehensive view of the individual and his/her needs where coordinated interventions from medical, pedagogical, psychological and social aspects of the functional limitations are taken into consideration [51].

The family-centered approach has had an influence on health services and habilitation over the last few decades. The approach is based on the notion that parents know their children best, that all families are unique and that children function optimally in supportive families and communities. Therapists are considered as collaborators, and together with child and family treatment goals are identified [52].

The prevention program, CPUP was started in Skåne and Blekinge in 1994, through cooperation between the Orthopaedic Departments and the Habilitation Centers to

prevent the development of hip dislocation and development of severe contractures in children and youth with CP. Since 2005 the whole of Sweden was included. A program was constructed where all children with CP are examined by standardized regular measurements by physiotherapists and occupational therapists, combined with regular radiographic screening of the hips and spine based on GMFCS levels and other examinations. Early signs of musculoskeletal deformities can be identified and relevant treatment organized. Children in need of additional spasticity reduction are identified and referred to the spasticity team at the specialist clinic. The prevention program has shown to prevent hip dislocation and the development of severe contractures in a total population with CP [53].

Children with severe spasticity interfering with function can be referred to the spasticity team at the specialist clinic at from the local habilitation centre, as a recommendation from CPUP or by neuropediatricians or orthopaedic surgeons seeing children with CP at the local habilitation centers. The choice of spasticity reducing methods should to be done by a multi-disciplined team with experience of selection, treatment and evaluation of treatment effects of the different options as well as the combinations of options[54].

The spasticity team at the specialist hospital consists of neuropediatricians, pediatric orthopaedic surgeons, neurosurgeons, specialized physiotherapists and on requests hand surgeons, occupational therapists and certified prosthetist and orthotists, To fully understand how the muscle tone affects the child, it is important to include the child and family in the team as well as representatives from the local habilitation, and together set realistic goals for the interventions [54]. The spasticity reduction is regarded as a tool to gain new prerequisites to perform physical training and muscle stretching. Physiotherapists have an important role in the process of recommending different treatment options for the children, discussing realistic goals with child and family, performing assessments of function prior to interventions, providing different mobility aids, recommending orthoses and standing frames to enhance joint stability and weight bearing, provide individually targeted motor training and performing follow-up of functional outcome after different interventions.

Treatment of spasticity

At present, there is no treatment that will repair existing damage to the brain structures that control muscle coordination and movement. However, several interventions are available to reduce excessive muscle tone and to decrease the impact on daily activities and to improve functional performance in children with CP [55]. Spasticity treatment can be divided into three principal groups according to how and

where they affect the spasticity; treatment affecting the peripheral structures, increasing spinal inhibition and decreasing spinal excitation.

Treatment directed towards the peripheral structures

The purpose of *physiotherapy* treatment methods is to improve or maintain functional levels, increase or improve the repertoire of motor skills and to minimize contractures and deformities [56]. To affect muscle tone, following neurophysiological principles have traditionally been used, but research evidence to support the efficacy of treatments is often lacking. Sensory stimulation techniques (e.g. vibration, ice and approximation), biomechanical approaches involving altering muscle length (e.g. stretching, use of casts splints and orthoses) and the alteration of the position of a patient may affect muscle tone [22]. Medical, surgical and/or orthotic or casting treatment options to treat spasticity have been recommended to be combined with physiotherapy interventions to optimize the spasticity reducing effects [54].

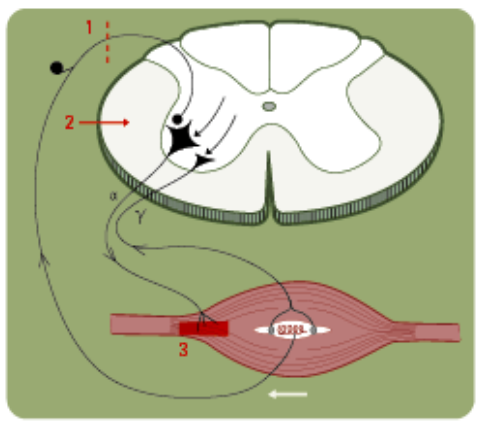

Figure 1.
Schematic overview of the locations where different spasticity treatment act; 1- Selective Dorsal Rhizotomy- sensory input is reduced by sectioning dorsal rootlets, 2- Baclofen- reduces the stretch-reflex and other poly-synaptic reflexes by inhibiting the release of excitatory neurotransmitters , 3- Botulinum toxin- toxical denervation of motor end plates. Figure is used by permission from Läkartidningen.

Selective Dorsal Rhizotomy (SDR) has been used to permanently reduce moderate to severe lower extremity spasticity and improve function in children with spastic diplegia [57]. By cutting a proportion of dorsal (sensory) rootlets, SDR reduces the force in the spinal stretch reflex (Figure 1). Electromyography (EMG) is used to indentify spinal roots and, together with the functional status, to guide the decision of which dorsal rootlets to cut. Rootlets between the spinal levels S2- L2 are identified

and stimulated. Dorsal rootlets with the most pathological responses are cut. Different centres report between 30-65% of cut rootlets [58-62].

As the ultimate goal of the intervention is to improve function, it should always be combined with physiotherapy. The operation is recommended for young children with spastic diplegia, without dyskinesia or ataxia, without significant cognitive disability and with walking capacity or ambulation within reach [60-63].

SDR combined with physiotherapy has been proven to have beneficial impact on the components of body function and structure according to the International Classification of Functioning, Disability and Health (ICF) [64]. Lower limb spasticity is consistently relieved and lower limb active and passive range of motion have been shown to increase [60-62, 65-67]. Strength and gait velocity are improved [68, 69]. Evidence for positive impact in the component of activity and participation according to the ICF has been reported [61, 70-72]. Improvements in activities of gross motor function measured by the Gross Motor Function Measure (GMFM) have been shown [70]. Performance in functional skills and mobility, as measured by the Pediatric Evaluation Disability Inventory (PEDI), was improved [71]. Meta-analysis data derived from three randomized studies concluded that SDR in combination with physiotherapy treatment had a positive effect on gross motor function one year after the operation. The children who had undergone this treatment had a greater functional improvement than children only receiving physiotherapy. A direct relationship between the percentage of dorsal rootlets transected and functional improvement was found [73]. Lasting functional improvements in gait have been shown 10 and 20 years after SDR [74, 75].

SDR may reduce the need for orthopaedic procedures in patients with spastic CP [76, 77] especially if the SDR operation was conducted in children before the age of five years [78, 79]. SDR has been suggested to have a positive effect on the prognosis of hip dislocation [80]. Perioperative complications are rare [55]. A high incidence of spinal deformities has been reported after SDR [81-84]. However, Langerak et al. found no significant increase of spinal deformities in a 16-27 year follow-up after SDR [85].

Children with spastic cerebral palsy may require *orthopaedic surgery* to prevent, or correct secondary deformities obtained by muscle over-activity and imbalance. Uncorrected deformities may cause pain, restrict functional ability or care and result in joint subluxations or dislocations. Correction typically involves tenotomies, muscle transpositions and osteotomies [86]. *Serial casting,* to lengthen shortened muscles and soft tissues, or to correct contractures, has proven to be effective, and is often used in combination with btx injections [87].

Orthoses are designed to provide joint stability, to hold the joint in a functional position and/or to keep tight muscles stretched. Ankle-foot orthoses (AFOs) are most

commonly used in CP to reduce dynamic equinus [86]. Even if there is limited evidence to support physiological mechanisms by which tone reduction may arise using orthoses, the techniques are widely used [88]. Orthoses are used to prevent or treat secondary effects of spasticity; restriction in range of motion and joint deformities. The use of standing frames for weight bearing has been shown to increase the range of motion in popliteal angle in non-ambulant children [89] and decrease lateralization of the hips in bilateral spastic CP [90].

Botulinum toxin (btx) targets the synaptic vesicle fusion mechanism at the neuromuscular junction. A reversible denervation occurs because vesicles cannot fuse with the synaptic membrane and acetylcholine cannot be released, which causes muscle weakness (Figure 1). At effective doses the duration of clinical action is three months; however this can vary between children and within the same child between injections [86]. A Cochrane review of the treatment effects of btx found no strong controlled evidence to support or refute the use of btx for the treatment of leg spasticity in cerebral palsy. The report was updated in 2009 with no changes in conclusions [91].

Phenol and *alcohol* are non-selective proteolytic agents which cause denervation where they are injected. The denervation causes muscular weakness and occurs only a few millimeters from the injection site. The effect lasts for 3-6 months in alcohol injections and 4-8 months for phenol. Side effects such as pain, possible muscle fibrosis and dysesthesias lasting for several weeks, are probably why these methods are not frequently used in children [92].

Treatment to increase spinal inhibition

Administering anti spastic agents orally has the advantage of being easy to distribute, but with the disadvantage of systemic effects and unwanted side effects. Most trials have been carried out on adults and very few on children. Common medications given orally are *Baclofen* and *Diazepam* [86]. Oral baclofen in therapeutic doses reduces spasticity mildly but may also be associated with decreased concentration or lethargy [93] (Figure 1). Diazepam is a benzodiazepine which is a cheap, well tolerated and effective muscle relaxant. However, drowsiness limits its use. Significant reduction of hypertonia, improvement in the passive range of motion and increased active movements in mainly quadriplegic children receiving diazepam has been reported in a double blind placebo controlled study [94].

By administering baclofen to the intrathecal space via a catheter attached to an implanted infusion pump, a significant dose reduction can be achieved. Less than 1% of the orally delivered dose is enough for spasticity reduction via the pump (Figure 1).The dose is distributed within the cerebrospinal fluid and migrates into the superficial layers of the spinal cord [95]. *Intrathecal baclofen* (ITB) has been shown to decrease muscle tone and improve functional outcomes in children with CP [96].

Treatment to decrease spinal excitation

Tizanidine and *clonidine* are distributed orally and diminish the release of excitatory amino acids from pre synaptic terminals of spinal inter neurons. This is considered to decrease tonic stretch reflex and the degree of antagonistic co-activation. It has been shown to more effectively potentiate the effects of intramuscular Btx injections in gastrocnemius than baclofen in children with CP [97].

Spasticity treatment options over time

When SDR was introduced in Sweden in 1993, the available spasticity treatment options were limited; neither btx nor ITB were available for this group of children. SDR, orthopaedic surgery, casting and physiotherapy treatment were the available options. Today btx, combined with physiotherapy treatment, is the first choice for spasticity treatment in young children. ITB is also an available option, especially for children in GMFCS IV-V. The numbers of children undergoing SDR are today reduced compared to the early years, probably because other treatment options are now available.

Theoretical framework for function

In 2001 the World Health Organization presented the International Classification of Functioning Disability and Health (ICF) [64]. This is the international standard for conceptualising health and disability of people and populations and for collecting and coding data on function. The aim of the ICF is to establish a common language and framework to describe functioning and health, to provide a scientific basis for understanding and studying health and health-related determinants, to permit comparisons of data and to provide a systematic coding scheme.

The International Classification of Functioning Disability and Health for Children and Youth (ICF-CY) is derived from the ICF and is designed to record the characteristics of the developing child and its influence on its surrounding [1]. The classification builds on the conceptual framework of the ICF. It uses a common language and terminology for recording problems involving body function and structures, activity limitations and participation restrictions seen during infancy, childhood and adolescence. Environmental factors relevant to the individual are also recorded [1]. The ICF-CY has been used in this thesis as a framework of functional outcome.

The ICF-CY has different parts, divided into components:

Functioning and Disability consists of Body Functions and Body Structures and Activities and Participation. Body functions are defined as “the physiological

functions of body systems (including psychological functions) and body structures as "anatomical parts of the body such as organs, limbs and their components". Activity "is the execution of a task or action by an individual" and participation "is involvement in a life situation". The negative aspects of functioning and disability are impairment, activity limitations and participation restrictions. Impairment is defined as "problems in body function or structure such as significant deviation or loss". Activity limitations are defined as "difficulties an individual may have in executing activities" and participation restrictions are "problems an individual may experience in involvement in life situations [1, p. 9-12]

Contextual factors consist of Environmental Factors and Personal factors. Environmental factors are either individual or societal and are defined as factors making "up the physical, social and attitudinal environment in which people live and conduct their lives". Positive factors are facilitators and negative factors are barriers. Personal factors "are the particular background of an individual's life, and comprise features of that individual that are not part of a health condition or health status" [1 p. 15].

In order to visualize the interaction between the various components, see Figure 2. The individual's function in a specific domain represents an interaction between the health condition and environmental and personal factors. An intervention in one component may affect one or more components. The interaction works in two directions and the health condition itself can be affected by the presence of disability [1].

The framework of the ICF can be used to consider the effects of therapy from an overall perspective of children's functioning. It may help the clinician to identify the component level of assessment findings, clinical concerns, client goals and outcome measures.

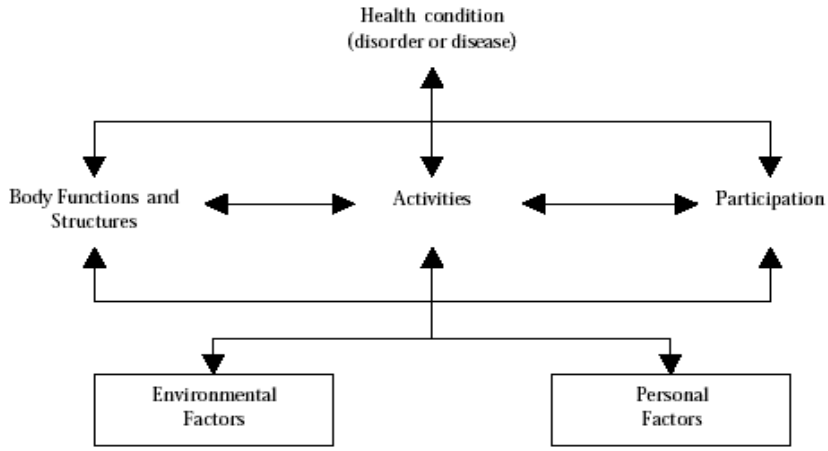

Figure 2.
Interactions between the components of the International Classification of Functioning, Disability and Health (ICF). Reprinted with permission from the WHO.

Measurements of function in children undergoing SDR

Treatment of spasticity in children undergoing SDR has the possibility to influence several components of the ICF-CY. Measurements of functional outcomes should therefore be performed using standardized classifications and measures all over the ICF-CY components body functions and body structures, activities and participation (Figure 2) [1].

Assessments of body functions and body structures

To assess increased *muscle tone in children* with CP, the passive resistance to the lengthening of a muscle can be rated according to different scales. The most commonly used scales are the Ashworth scales rating the dynamic component, and the Tardieu scales rating both the dynamic and the passive component. Both the Ashworth and the Tardieu scales are available in many modifications [98-103].

The degree of *deep tendon reflex* response can be rated according to a 5-point scale, namely the National Institute of Neurological Disorders and Stroke (NINDS) scale [104].

The maximal *passive range of motion* can be measured with the use of a goniometer and standardised anatomical landmarks and methods often recommended by the American Academy of Orthopaedic Surgeons [105].

Selective voluntary motor control (SVMC) describes the performance of specific isolated joint movements upon request, as opposed to the habitual activation of selected muscles during functional tasks. The Selective Control Assessment of the Lower Extremity (SCALE) is a clinical tool developed to quantify SVMC in patients with CP [106].

To monitor development of gait pattern over time, *three- dimensional gait analysis* (3DGA) may be used. Outcome studies have generally reported selected univariate gait variables such as temporo-spatial parameters, joint excursions and angular velocities. However, separate univariate parameters cannot fully capture the overall picture due to the complex correlation among separate variables. By using multivariate statistical methods e.g. principal component analysis the correlation between gait variables can be determined [107]. The recently introduced multivariate overall score Gait Profile Score (GPS) and Movement Assessment Profile (MAP) obtained from three-dimensional gait analysis (3DGA) data selected from principal component analysis, offer a means to monitor gait pattern deviation over time [108].

Assessments of activities and participation

The main symptom of CP in children is restriction of *motor function.* The severity of functional limitations can be classified according to a five level ordinal scale using the observational classification system, Gross Motor Function Classification System (GMFCS), introduced in 1997 [109] and the expanded and revised version *(GMFCS E&R)* from 2007 [110]. The GMFCS is a classification system used to describe and classify functional ability in children with CP [109]. It is based on gross motor development of self-initiated movements with emphasis on sitting and walking. Children with spastic diplegia are a clinically heterogeneous group with limitations of gross motor function spanning from minor to severe. When evaluating functional outcomes for children with spastic diplegia their different prerequisites for change must be considered. The GMFCS provides a possibility to distinguish between different levels of functional limitations enabling comparison between children with similar prerequisites. For the description of the GMFCS levels for children aged 4-6 years, see Table 1.

The Gross Motor Function Measure (GMFM) is a measure commonly used for assessing a child's gross motor function capacity in a standardized observational way [111]. The GMFM is a criterion- referenced measure based on normal gross motor developmental mile stones; all items are achievable by a five-year old child without any motor disability. It was designed to yield an index of gross motor function, enabling changes in function to be evaluated after interventions, or monitored over time for children with CP. The GMFM is available in both an 88-item and a 66-item version. The scoring options can be used for all CP subtypes, however the slightly different psychometric properties can be further evaluated.

Table 1.
Functional abilities for children aged 4-6 years according to the Gross Motor Function Classification System (GMFCS). For a complete description of the GMFCS see Palisano et al. [109].

GMFCS level	Description of gross motor function
I	Children floor sit with both hands free to manipulate objects. Movements in and out of floor sitting and standing are performed without adult assistance. Children walk as the preferred method of mobility without the need for any assistive mobility device.
II	Children floor sit but may have difficulty with balance when both hands are free to manipulate objects. Movements in and out of sitting are performed without adult assistance. Children pull to stand on a stable surface. Children crawl on hands and knees with a reciprocal pattern, cruise holding onto furniture and walk using an assistive mobility device as preferred methods of mobility.
III	Children maintain floor sitting, often by "W-sitting" (sitting between flexed and internally rotated hips and knees), and may require adult assistance to assume sitting. Children creep on their stomach or crawl on hands and knees (often without reciprocal leg movements) as their primary methods of self-mobility. Children may pull to stand on a stable surface and cruise short distances. Children may walk short distances indoors using a hand-held mobility device (walker) and adult assistance for steering and turning.
IV	Children floor sit when placed, but are unable to maintain alignment and balance without use of their hands for support. Children frequently require adaptive equipment for sitting and standing. Self-mobility for short distances (within a room) is achieved through rolling, creeping on stomach, or crawling on hands and knees without reciprocal leg movement.
V	Physical impairments restrict voluntary control of movement and the ability to maintain antigravity head and trunk postures. All areas of motor function are limited. Functional limitations in sitting and standing are not fully compensated for through the use of adaptive equipment and assistive technology. At Level V, children have no means of independent movement and are transported. Some children achieve self-mobility using a powered wheelchair with extensive adaptations.

Gross motor function measured by the 66 item version, GMFM-66 has been shown to develop over time, according to age and GMFCS levels, in 657 children participating in the Ontario Motor Growth (OMG) study [112].

Reference percentiles were constructed by selecting a clinically appropriate comparison group and developing a statistical summary of the distribution of the scores of the group between the ages of 2-12 years. The GMFM-66 percentiles measure relative ability compared to other children of the same age and GMFCS levels. A peak in GMFM-66 scores in children between 7-8 years of age, whilst a decline in scores for children in GMFCS levels III, IV and V have been shown during early teen ages (Figure 3) [113]. Presently, percentile scores are not available after the age of 12 years.

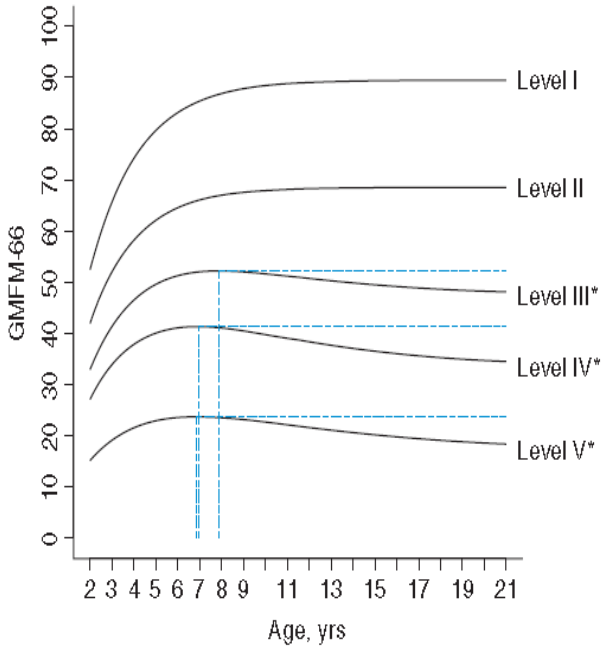

Figure 3.
Predicted Gross Motor Function Measure (GMFM-66) motor scores as a function of age by Gross Motor Function Classification System (GMFCS) level. * GMFCS levels with significant peak and decline. Dashed lines illustrate age and score at peak GMFM-66 score. Figure is used by permission of John Wiley & Sons.

Pediatric Evaluation of Disability Inventory (PEDI) is a generic standardized questionnaire for the multidisciplinary team for evaluating *functional performance*, program monitoring, documentation of functional development and clinical decision-making [114]. Emphasis is placed on the child's performance in everyday activities, not best capacity in a single test situation.

The Functional Mobility Scale (FMS) can be used in order to classify *performance of functional mobility* in children with CP and to document changes over time in the same child [115]. The FMS is distributed as a questionnaire and classifies functional mobility in children, taking into account the range of assistive devices a child might use. The FMS is rated according a six level ordinal scale in three different distances; 5, 50 and 500 meters, representing the everyday contexts; home, school and community.

Psychometric properties of measures

When selecting outcome measures for clinical practice or research the decision needs to be based on a critical evaluation of the evidence of validity and reliability [116].

Validity refers to the degree of which the test measures what it is intended to measure. *Face validity* refers to whether the test appears to be meaningful to tester and patient. *Content validity* refers to how well the test samples the phenomenon under study. *Construct validity* is the extent to which predefined hypothesized associations among different measures of the concept are confirmed [5]. *Longitudinal construct validity* is the extent to which the instrument can measure change in the concept in which the measure is supposed to measure [9].

Responsiveness is defined as the ability of an instrument to measure a meaningful or clinically important change in a clinical state [9]. *Sensitivity to change* is the ability of an instrument to measure change in a state regardless of whether it is relevant or meaningful to the decision maker [9].

Reliability reflects the amount of error, random and systematic, inherent in any measurement. *Inter-rater reliability* is the consistency of measurements between raters. *Intra-rater reliability* is the consistency of measurements between measurements recorded by the same rater [5].

Psychometric properties of the measures used in this thesis are reported in the method section.

Physiotherapy treatment approaches in children with CP

The goal of physiotherapy treatment is to improve quality of life for the children and their families, to optimize their participation in daily activities and to prepare for improved quality of life during adulthood[117].

Today, the two most common approaches within the field of physiotherapy for children with CP are the task/context- focused and the child- focused approaches.

In the *task/context focused* approach the goal of treatment for the child is to achieve a specific functional goal which has been identified together with child, family and therapist. Emphasis is placed on the success of the task, rather than on obtaining normal patterns of movement. Different contexts may need different solutions to the problem. This approach has its theoretical basis in the dynamic systems theory, where

movement is always goal-oriented and context specific [118]. It is most likely that treatment will have a good effect when it is presented at a time when the child is trying him/herself to do a new task or to change performance of an established task. This transition period is a window of opportunity, known in the dynamic systems theory as the period when the movement patterns are more easily disturbed and the child is most ready to achieve a new goal. The treatment will be planned based on both different constraints and enablers of the specific goal identified by the child, family and therapist. The practise of the functional goal is best performed in the most appropriate environment, namely the natural setting.

The term, top-down is used for this approach where first functional goals, and then specific constraints are identified [119, 120].

The *child-focused approach* concentrates on remediation of body function and structure and is often known as a neuro- maturational approach. This approach is significantly influenced by hierarchical theories of motor development. In children with CP, this is exemplified by improving/maintaining the range of motion through stretching and casting, strength training and the facilitation of normal movement patterns, etc. Improved functional performances outcomes obtained by changes in body function and structure.

This is known as the bottom- up approach, as the process involves identifying first the impairments and then the functional limitations [119, 120].

Even if top-down and bottom-up approaches are theoretically opposed to each other, a combined approach is perhaps to be recommended. Depending on individual, environmental and activity factors, the most appropriate approach for each child may differ [119].

Rationale of the thesis

There is a lack of longitudinal studies of functional outcome after SDR combined with physiotherapy. As the intervention causes permanent spasticity reduction, it is of great importance for the future selection of suitable candidates, to identify how children respond to the intervention. There is a need of valid and reliable measures for evaluation of treatment in the components of body functions, body structures, activities and participation according to the ICF-CY for a better understanding of the impact of procedures [1].

Aims of the thesis

The overall aim of the thesis was to

- describe the development of children's function over a period of 10 years following SDR combined with physiotherapy.

The specific aims of the different studies were to

- evaluate long-term functional outcomes, safety and side effects during five years postoperatively in 35 children undergoing SDR combined with physiotherapy (Paper I).

- analyze the panorama of orthopaedic surgery in children five years after SDR (Paper II).

- study longitudinal construct validity of GMFM-88 total, goal total score and GMFM-66 over five years among children with CP undergoing SDR (Paper III).

- explore the utility of the Gait Profile Score (GPS) and the Movement Assessment Profile (MAP) in a long-term follow-up in seven children undergoing SDR (Paper IV).

- describe changes in muscle tone, passive range of motion and capacity of gross motor function between pre- and 10 years postoperatively in children following SDR combined with physiotherapy in relation to GMFCS levels and age (Paper V).

- identify factors that could explain changes in muscle tone, passive range of motion between pre- and 10 years postoperatively (Paper V).

- describe performance of mobility at 10 years after SDR (Paper V).

Methods

Participants

All participants had spastic diplegia and were undergoing SDR in combination with physiotherapy treatment at Lund University Hospital between 1993 and 2004. Children eligible for SDR were below seven years of age, had a diagnosis of spastic diplegia, had spasticity interfering with present and future motor function and daily activities and had a willingness to receive pre- and postoperative physiotherapy treatment. Contraindications were the presence of dystonia, ataxia and fixed contractures or earlier major orthopaedic surgery [57].

In Paper I and II, the first 35 consecutively selected and SDR operated children were included (Table 2). Mean age (median, range) at SDR for the children was 4.5 (4.3, 2.5-6.6) years. In Paper I subgroups according to preoperative GMFCS levels were created; GMFCS I-II (n=9), III (n=10) and IV-V (n=16). In Paper I results were also analyzed for the group as a whole; GMFCS I-V (n=35).

The first consecutively selected and SDR operated 41 children were included in Paper III (Table 2). Mean age (median, range) at SDR was 4.4 (4.2, 2.5-6.6) years. Two subgroups according to preoperative GMFCS levels were created; GMFCS I-III (n=23) and GMFCS IV-V (n=18), results was also analyzed for the group as a whole GMFCS I-V (n=41).

In Paper IV, all seven children in GMFCS I and II who walked without walking aids prior to SDR and had been undergoing 3DGA during five postoperative years in SDR follow-up were included (Table 2). Mean age (median, range) at SDR was 5.3 years (6.3, 3.8-7.2).

In Paper V, the first 35 consecutively selected and SDR operated children were invited to participate, 29 of which gave their informed consent (Table 2). Mean age (median, range) at 10 year follow-up was 14.8 (15.0, 12.8-17.1) years. All 29 were previously also included in study I, II and III. In Paper V, subgroups according to preoperative GMFCS levels were created; GMFCS I-II (n=8), III (n=8) and IV-V (n=13).

Data from one child was included in all five studies (Figure 4). For distribution according to GMFCS levels, see Table 2.

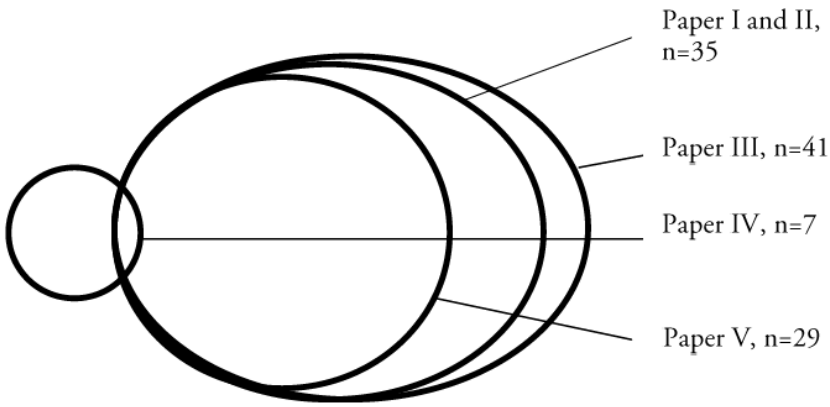

Figure 4.
Participants overlap between studies.

Table 2.
Participants according to preoperative levels of GMFCS.

	Paper I	Paper II	Paper III	Paper IV	Paper V
GMFCS I	1	1	1	2	1
GMFCS II	8	8	9	5	8
GMFCS III	10	10	13	-	7
GMFCS IV	15	15	17	-	12
GMFCS V	1	1	1	-	1
Total	**35**	**35**	**41**	**7**	**29**

Selective Dorsal Rhizotomy (SDR) and physiotherapy

The spasticity team at the specialist clinic collaborate closely when selecting children for SDR. The actual status of the child including any additional disorders or impairments, are investigated by the neuropediatrician to ensure that the child has the appropriate medical background. Neuroimaging has been used especially during the last decade, to map brain morphology. Children are seen at repeated visits at the specialist clinic to follow the development of the child and to make sure that inclusion criteria are met before a recommendation of SDR or not are made.

The pediatric physiotherapist assesses voluntary strength, consequences of spasticity in PROM, gross motor function and activities in daily life. Personal and environmental factors are also important to consider in the selection process and should be identified and considered. These factors may be the child's own motivation to move and play and the support from family and the cooperating local habilitation team. The knowledge of these factors is based on experience from previously SDR operated children followed by the spasticity team at the specialist clinic during many years. However, for the children and families undergoing SDR, life events of different magnitude occur over times which are not possible to predict but may highly influence development of functional outcome. Continuity in the spasticity team at the specialist clinic, where the body of experience is growing throughout the years, is considered to be of great importance for a successful selection of future SDR candidates.

Long- and short term goals are thoroughly discussed with the child, family and local habilitation team. It is also important to discuss prior to the intervention what happens when spasticity is reduced and in what situations the spasticity even may be useful e.g. in weight-bearing, standing, walking and in transfers [58].

During the operation dorsal rootlets between S2 to L2 are accessed by a block laminoplasty between L1-L5 and an incision of the dura to expose the cauda equina. Each root is identified and separated into smaller units. EMG recordings are obtained from the hip adductor, vastus lateralis, tibialis anterior, hamstrings and gastrocnemius muscles of both sides and in the external anal sphincter. Each rootlet unit is stimulated with a handheld cauterizing forceps connected to a current stimulator and the EMG response is recorded for the muscles. All rootlets in each level are stimulated before deciding which to cut and which to spare. The neurophysiologist is informed of the preoperative status in order to dose the spasticity reduction throughout the spinal rootlets. The number of rootlets cut is determined from the occurrence of

pathological responses in combination with preoperative assessments of spasticity interfering with motor function. The same neurosurgeon (L-G Strömblad) and neurophysiologist (G Andersson) performed all the SDR operations in all children included in Papers I-V.

Postoperative physiotherapy is started at the fifth postoperative day and gradually increased during the first month. The first ten days as an inpatient at the Neurosurgical intensive care unit and the Children's hospital and the last two weeks as an inpatient at a regional habilitation unit for continued postoperative physiotherapy in a more home-like environment. Physiotherapy is incorporated into the child's daily activities, promoting functional skills in playing, dressing, grooming, transfers and mobility. Weight bearing is introduced early postoperatively and is gradually increased. Standing shell is recommended for all children one to two hours a day, to promote symmetrical load for the benefit of skeletal growth, maintaining or improving muscle length by long duration stretching, and for postural control and improving balance reactions in an optimal standing position. Also the use of arms and upper body may be improved when standing in a standing shell. As soon as the scar is healed, hydrotherapy is introduced. After discharge, the physiotherapists at the local habilitation continues to implement functional activities and participation according to preoperatively set goals in daily activities in close collaboration with the child, family and the physiotherapist in the spasticity team at the specialist clinic to optimize functional outcome.

The recommended frequency of individualized treatment sessions is one hour, twice weekly, during the first six months, and once a week during the following 18 months. Additionally physical leisure activities are encouraged.

During rehabilitation, the physiotherapist at the specialist clinic coaches the child, family and physiotherapist at the local habilitation by regular re-visits with measurements of function monitoring the development of function. Based on these outcomes, long-term goals for the individual child and experience from previous similar patients, an individual physiotherapy treatment plan is drawn up and short-term goals revised.

Regular follow-up visits at the spasticity clinic are performed after 3, 6, 12 and18 months as well as 3 and 5 years postoperatively, support the rehabilitation process. The ten year follow-up was introduced as optional in 2003.

Table 3.
Study design, postoperative follow-up time after Selective Dorsal Rhizotomy (SDR), classifications and measures used in Papers I-V.

	Paper I	**Paper II**	**Paper III**	**Paper IV**	**Paper V**
Study design	Follow-up study	Follow-up study	Measurement study	Measurement study	Follow-up study
Follow-up time after SDR	6, 12, 18 months, 3 and 5 years postoperatively	5 years postoperatively	6, 12, 18 months, 3 and 5 years postoperatively	1, 3 and 5 years postoperatively	10 years postoperatively
Modified Ashworth	X				X
Reflexes and clonus	X				
PROM	X				X
3DGA				X	
GMFCS	X	X	X	X	X
GMFCS-E&R					X
GMFM-88 total score	X		X		
GMFM-88 goal total score	X		X		
GMFM-66	X		X	X	X
PEDI	X				
FMS					X

Gross Motor Function Classification System (GMFCS), Gross Motor Function Classification System- Expanded and Revised (GMFCS-E&R), passive range of motion (PROM), Three dimensional gait analysis (3DGA), Gross Motor Function Measure (GMFM), Pediatric Evaluation, Disability Inventory (PEDI), Functional Mobility Scale (FMS).

Assessments and measurements

To monitor the development of functional outcomes, the following assessments and measures were used prior to SDR and at follow-ups (Table 3). Functional outcome measures used in the thesis are presented according to the ICF model, see Table 4.

Table 4.
A schematic overview of outcome measures used in this thesis in relation to the International Classification of Functioning, Disability and Health- Children and Youth version (ICF-CY) [1].

Health condition **Cerebral palsy**				
Functioning & disability			**Contextual factors**	
Body functions & Body structures	**Activity & Participation**		**Environmental factors**	**Personal factors**
	Capacity	**Performance**		
Reflexes PROM Modified Ashworth scale 3DGA	GMFM	GMFCS GMFCS-E&R PEDI FMS	GMFCS GMFCS-E&R PEDI FMS	n.a.

Passive Range of Motion (PROM), three-dimensional gait analysis (3DGA), Gross Motor Function Measure (GMFM), Pediatric Evaluation Disability Inventory (PEDI), Functional Mobility Scale (FMS), not applicable (n.a.)

Body function and body structure

Muscle tone in hip adductors, hip flexors, knee flexors and plantarflexors were assessed by the same two experienced physiotherapists A Lundkvist Josenby and E Nordmark (ALJ and EN) according to the Modified Ashworth scale modified by Peacock and

Staudt [98] (Table 5) (Papers I and V). Reliability studies for the Ashworth scale modified by Peacock and Staudt have not been published.

Table 5.
Modified Ashworth scale by Peacock and Staudt [98].

Score	Grade	Definition
0	Hypotonic	Muscle tone is less than normal.
1	Normal	No increase in muscle tone.
2	Mild	Slight increase in tone; "catch" or minimal resistance to movement is felt during passive movement throughout less than half of the range of movement.
3	Moderate	Marked increase of muscle tone; resistance to movement is felt during passive movement through more than half of the range of movement. However, passive movement is easily performed.
4	Severe	Considerable increase in muscle tone, passive movement is difficult to perform.
5	Extreme	Affected part is rigid in flexion or extension.

Deep tendon reflexes and clonus in achilles, adductor and quadriceps tendons at the pre- and postoperative follow-ups were examined by the two neuropaediatricians J Lagergren or L Westbom (Paper I). The degree of deep tendon reflexes response was rated in a 5-point scale, the National Institute of Neurological Disorders and Stroke scale (NINDS) [104]. Clonus was arbitrarily graded into a three point scale: no clonus, 1-6 beats, and $\geq$ 7 beats.

Passive Range of Motion (PROM) was measured for hip abduction with the hips and knees extended, popliteal angle with hips flexed 90° with knees maximum extended, and ankle dorsiflexion with the knee extended and the foot inverted (Papers I and V). Variability of measurements in assessments of PROM using a goniometer in children with spastic diplegia have been shown to vary between five and 15° between measurements in the major joints of the lower limbs, with wide margins of error [99, 102, 121]. The physiotherapists (ALJ and EN) performed all the measurements.

Three dimensional gait analysis (3DGA) was performed at the Motion Laboratory Scandinavian Orthopaedic Laboratory, University Hospital in Lund, Sweden (Paper IV). Marker position data were captured by a Vicon 512 (Vicon Motion System, Ltd, Oxford, UK) which is a 3D passive marker motion capture system. It consists of six cameras with a sampling frequency of 100 Hz, one data station and one computer in which the information is gathered and processed. Spherical reflective surface markers were placed according to the Helen Hays marker protocol on the lower limb [122]. Kinematic data were derived from a plug-in-gait model based on the Newington-Helen Hays model [123].

In the review article by McGinley et al. it was concluded that in 3DGA, the highest reliability was found in the hip and knee in the sagittal plane. The least errors were found in pelvic rotation, pelvic obliquity and hip abduction. The highest errors occur in the transverse plane in hip and knee, of which hip rotation has been reported to have the largest measurement error. In regular 3DGA measurement errors of two degrees or less are considered as acceptable. Errors above 5° should raise some concerns as they may mislead the clinical interpretation [124].

Data was collected by ALJ and the staff at the motion laboratory at pre- and 1, 3 and 5 years postoperatively.

The children were instructed to walk at a self selected pace along a 10 m marked walkway. Marker data were collected over the middle three meters of the walkway where a force plate collected data from the stance phase of each leg. The children were always barefooted when assessed.

In the *Gait Profile Score (GPS)* and the *Movement Assessment Profile (MAP)* nine kinematic parameters are included; pelvic -tilt, -obliquity, -rotation; hip-flexion, -adduction, -rotation; knee flexion; ankle dorsiflexion and foot progression (Paper IV). Bilateral data was collected for all parameters except for the pelvis where only data from the left side was used as the pelvis is considered as a single segment moving as one unit.

Mathematically both the GPS and the MAP were calculated as the root mean square difference between the data and the average of a reference data set and are reported in degrees. The GPS is a single score calculated as the root mean square average score of the nine kinematic parameters. The GPS can be obtained as an overall score or scores from the left and right side. The MAP is reported as a bar graph and figures consisting of nine relevant variables for the right and left side. A result of zero degrees corresponds to normal gait pattern. The higher the scores of the GPS and the MAP, the more deviation from normal gait is seen [108].

The GPS have been tested for intra-session variability and the median Inter Quartile Range (IQR) was 0.67°. Six percent of the studied group showed larger IQR than two degrees [108].

The reference data set consisted of 12 typically developing children with a mean age (SD, median, range) of 8.6 (1.6, 8.5, 7-12) years.

The first three trials containing kinematic and kinetic data for each child and side were included, when available, at pre-, 1 year, 3 and 5 years postoperatively. Three trials per side and child were included in calculations of GPS. All selected 3DGA data were batched and uploaded to Gaitabase by ALJ according to the instructions provided in the manual [125].

Families were questioned about the *child's general health* including appetite and sleep, micturition and bowel habits, epilepsy, infections, pain and sensory disturbances such as hypo- or hyperestesisa or other health problems (Papers I, V). Weight and height were measured at all follow-ups and plotted into growth charts. Radiographs of the hip were performed prior to SDR and at least five years after and radiographs of the spine were performed prior to SDR and five and 10 years after SDR.

Activity and participation

The original *Gross Motor Function Classification System (GMFCS)* consists of a five level ordinal scale with definitions for four age bands; before the age of two years, between two and four years, between four and six years and between six and 12 years of age (Papers I- V). When the *Gross Motor Function Classification System- Expanded & Revised (GMFCS–E&R)* was published the age band for adolescents 12- 18 years of age became available and the age band 6- 12 years was modified (Paper V). More emphasis was placed on aspects of participation and personal choices in the age bands 6-12 years and 12-18 years [110]. Children whose gross motor function is classified as level I are as capable of motor activities as their typically developing peers but are limited in speed and agility, while children in GMFCS V have no means of independent mobility. In Table 1, the description of GMFCS levels for children ages 4-6 years is presented. The age band was selected, as it is in these ages most children are undergoing SDR in this thesis.

The validity of the GMFCS was established by nominal group process and Delphi survey consensus. Evidence of construct and content validity was obtained [109]. Construct validity was supported by high correlations between GMFM scores and GMFCS levels [126]. Inter-rater reliability was found to be κ=0.55 for children below the age of two and κ=0.75 for children between two to 12 years [109]. Inter-rater reliability between parents and health professionals was excellent [127]. The GMFCS have been shown to be stable in 73% of the children during childhood [128] and over the course of one year in 79% in parents report [129]. A systematic review in 2004 identified 102 citations and 75 journal articles that examined the psychometric properties or used it in research [130]. Together with GMFM-66 it can be used to

predict gross motor function based on data from a large group of all CP subtypes [112].

Swedish translations of the GMFCS and GMFCS-E&R were used [131, 132]. The gross motor function of the children was classified according to the GMFCS by ALJ and EN preoperatively and GMFCS-E&R by ALJ at 10 years postoperatively.

The original *Gross Motor Function Measure (GMFM)* contained 88 items scored in a four level ordinal scale with scoring instructions for each item defined in the manual [111] (Papers I, III). In GMFM-88 it is possible to obtain scores for five separate dimensions (A- Lying and rolling; B- Sitting; C- Crawling and kneeling, D- Standing and E- Walking, running and jumping). The dimension scores are calculated as a percentage of the maximum score for each dimension. The total score is the mean percentage score of all five dimensions. The goal total score is calculated for each child as the mean of the individually selected GMFM dimension scores. The dimensions for the goal total score were selected based on the child's functional status, age and areas of interest.

The GMFM-88 has been used in different settings by therapists worldwide. However, limitations in the measure and how it was used appeared [111]. The constructors used the Rasch-analysis to improve the scoring, interpretation and overall clinical and research utility [133].

The most recent version is known as the GMFM-66, as it contains 66 of the original 88 items (Papers I, III-V). The goal was to develop the GMFM-66 to be less vulnerable than the GMFM-88 to missing items and more responsive to children with both large and minor functional limitations. In the Rasch-analysis, 66 items were identified to contribute the most to the underlying construct of gross motor function. To improve reliability and validity, 22 items were deleted and an interval scale was created. Of the 22 items, 13 were from the Lying & Rolling dimension, five from the Sitting dimension and four items from the Crawling & Kneeling dimension. The GMFM-66 score is obtained by use of the Gross Motor Ability Estimator (GMAE) software [111]. The characteristics of the GMFM-88 and GMFM-66 are described in Paper III, Table 1.

The GMFM-88 total, GMFM-88 goal total and GMFM-66 scores range from zero to 100, the higher the score the better the function. Children with gross motor function classified as GMFCS level I, have generally higher GMFM scores than children of the same ages in e.g. GMFCS levels IV.

The original GMFM validation study included 111 children with CP, 25 children with acquired brain injury and 34 typically developing children who were tested twice over 5-7 months. Correlations between scores for change in motor function measured with the GMFM-88 and the judgments of change by parents, therapists and 'blind' evaluators supported the hypothesis that the instrument would be responsive to both

negative and positive changes [134]. Bjornson et al. studied 21 children with diplegia and quadriplegia and provided additional validation evidence of the responsiveness of the GMFM-88 [135]. The GMFM-88 was found to detect change in motor function with a mean increase of 4.2 points over six months in 24 infants with CP [136]. Russell et al. studied validity and responsiveness in 206 children with CP and found the mean change in motor function in GMFM-88 over six months to be 3.5 points [137]. The total and goal total scores in 18 children with CP who underwent SDR were found to respond to change in motor function over six and 12 months postoperatively, especially for children with milder impairment [72]. Vos-Vromans et al. studied responsiveness in GMFM-88 total and dimension scores over 18 months in children with CP aged 2–7 years. For total score ES was 0.6 and SRM 0.9 [138].

Both inter- and intra-rater reliability of the GMFM-88 have been reported to be good. Inter-rater reliability was found to be 0.77 and 0.88 at the first and second assessment respectively and intra-rater reliability 0.68 at the second assessment [139]. Bjornson et al. suggested that the GMFM was consistent in the measurement of gross motor skills. Children with CP exhibited stable gross motor skills during repeated measurement. Intra-class correlations (ICC) ranged from 0.76 to 1.00 [135].

GMFM-88 measurements in 537 children with CP performed by 110 physiotherapists were converted to GMFM-66 scores [140]. Children were excluded if they had had major interventions. Gross motor function in the 228 children reassessed after 12 months depended significantly on follow-up time since first assessment, age and severity of functional limitations. Other findings showed that children younger than five years changed more than older children, and less severely motor impaired children improved more than severely impaired children. They also found a high test-retest reliability and a similar result compared to GMFM-88 total score [140]. Test-retest and inter-rater reliability of ICC 0.97 and 0.98 respectively was found in 171 children with CP aged 0-3 years old [141].

Shi et al. explored the clinical consequences of deleting the 22 items from the GMFM-88 in children younger than three years [142]. They found the GMFM-66 responsive even for those young children who mainly have their functional abilities assessed in lying, rolling, sitting, crawling and kneeling positions. Wang and Yang compared the scoring options of the GMFM-88 total score and GMFM-66 to an external criterion of therapist's judgments of meaningful motor improvements after 3.5 months. They found the two scoring options to be equally responsive; however the GMFM-66 was found to have better specificity of therapist's judgments of meaningful motor improvements than the GMFM-88 [143].

Both ALJ and EN were trained and examined by the constructors in scoring the GMFM and were experienced in executing and scoring the test. The children were tested and videotaped by ALJ and EN preoperatively and at follow-ups after 6, 12, 18 months as well as 3, 5 and 10 years postoperatively.

The target group for *Pediatric Evaluation Disability Inventory* (PEDI) is children aged between 0.5 and 7.5 years (Paper I). It is also suitable for children older than 7.5 years if their functional ability is below that of non-disabled 7.5 year-olds. The PEDI questionnaire contains 197 items in three dimensions; functional skills, caregiver's assistance and modifications/adaptive equipment used. Each dimension has three domains: self-care, mobility and social function. Normative scores are available up to the age of 7.5 years and scaled scores can be used for all ages. PEDI scores range from zero to 100, the higher the score the better the functional performance.

The PEDI has been shown to be valid in children with and without disabilities [144]. Inter-rater reliability was investigated in a group of 30 children with disability between one to five years old by the constructors [114]. Excellent agreement in inter- and intra-rater reliability was shown [145]. Nordmark et al. compared the results of typically developing Swedish children aged 2-7 years, with the American normative data. They found the American normative data to be appropriate for reference purposes in Sweden [146].

The Swedish version of the PEDI was used in Paper I [147]. The Swedish questionnaire was available as a pilot version in 1994. The parents or caregivers were interviewed by the same physiotherapist (EN) preoperatively and during the follow-ups after 6, 12 and 18 months as well as after 3, 5 and 10 years.

Assessment of *Functional Mobility Scale* (FMS) was performed by asking the child or parent how the child actually ambulates, at home (representing the 5 m distance), at school (representing the 50 m distance) and in the community (representing the 500 m distance), and rating the type of mobility using a six point ordinal scale (Paper V). The different levels of the scale are: 1- uses wheelchair, 2- uses a walker or frame, 3- uses crutches, 4- uses sticks (one or two), 5- independent on level surfaces, 6- independent on all surfaces. The FMS is a performance measure, intended to rate what the child actually does, not what he/she can do, used to do or will be able to do [115].

The FMS has been found to be valid, reliable and sensitive to changes in children with CP of various levels of disability after orthopaedic surgery [115]. Harvey et al. found good agreement between children's performance and the parents' FMS reports [148].

The adolescents and their parents were interviewed regarding the functional mobility by ALJ or EN at 10 years postoperatively using a Swedish version of FMS [149].

Statistics

In Paper I Friedman's test was used to explore change over time for GMFM-88 total and goal total score, GMFM-66 scores and PEDI Scaled scores for Functional skills and Caregiver Assistance in Self-care and Mobility. Wilcoxon signed rank test was used to determine more specifically at what time during follow-up statistically significant changes in function appeared. Significance levels were set to $p = 0.01$ to correct for multiple comparisons. Results were analyzed for the group as a whole and for GMFCS subgroups.

In Paper III ES and SRM were used to evaluate longitudinal construct validity of the GMFM scoring options. ES was calculated as the mean difference between the baseline score and the follow-up scores divided by the standard deviation of the baseline score. SRM was calculated as the mean change score divided by the standard deviation of the change scores [6].

ES and SRM of 0.2–0.5 were classified as small, 0.5–0.8 as medium and > 0.8 as large [150].

Calculations of ES and SRM were performed between the measurements preoperatively and after 6, 12 and 18 months as well as 3 and 5 years postoperatively to study longitudinal construct validity of the GMFM scoring options as opposed to treatment effectiveness. Results were analysed for the group as a whole and for the two subgroups.

In Paper IV descriptive methods were used to illustrate changes in GPS and MAP for the seven children followed with 3DGA for five years.

In Paper V, effects on PROM, muscle tone and GMFM-66 10 years after operation of preoperative GMFCS level, baseline value, age at operation and birth year were estimated using ordinary linear regression with respect to preoperative GMFCS level. The linear mixed effects model were used for PROM and muscle tone. The mixed effects model was used in order to take into account the within-individual correlation of the outcome when including data on both limbs in the calculation. Selected level of statistical significance was $p \leq 0.05$.

The mean development of GMFM-66 scores according to age and preoperative GMFCS level was estimated using the statistical stable limit model, described previously in relation to GMFM-66 development by Hanna et al. 2009 [113].

The Statistical Package of Social Sciences (SPSS 15.0 and 17.0) was used for calculations in Paper I-IV. All analyses in Paper V were done using the statistical programming language R [151].

Ethics

According to Swedish National Board of Health and Welfare, clinicians are obliged to secure the quality of care by performing and reporting results of clinical studies in everyday practise. Approval from internal review boards is not required for this type of research. All participants have given their written informed consent to participate in the studies reported in Paper II, IV and V. Participants and all data have been handled according to the Helsinki convention. Approval was obtained by The Medical Ethics Committee at Lund University for Paper II (LU 414-02) and Paper V (LU 262-03) and at Linköping University for Paper II (Li 03-009).

Results

Long-term outcomes five years after selective dorsal rhizotomy (Paper I)

After SDR, the deep tendon reflexes decreased in the lower extremities ($p<0.001$). In most cases, they were completely extinguished. No further change occurred during the five years. Muscle tone in hip adductors, hamstrings and ankle plantarflexors decreased between preoperative and six months postoperative follow-ups and remained reduced over the five years ($p<0.001$).

The mean PROM increased for hip abduction, popliteal angle and ankle dorsiflexion for the group as a whole ($p<0.001$). The largest changes were detected at six months after SDR. Children in the GMFCS I-II showed statistically significant improvements for ankle dorsiflexion ($p=0.008$) and children in GMFCS III increased hip abduction ($p=0.009$). For children in GMFCS IV-V, statistically significant improvements were seen in hip abduction ($p=0.004$) and popliteal angle ($p=0.004$).

Increased lumbar lordosis was observed in four children at five years postoperatively. According to the radiographs three children had spondylolisthesis, of which one occasionally had back pain and the other two had no symptoms. Five children had developed scoliosis (Cobb angles 11-23°). None of the children had a brace or had undergone further spinal surgery. Preoperatively, 10 hips in seven children had a migration percentage (MP) > 33%. After five years, eight had improved and two had deteriorated, of which one had been referred to surgery to prevent hip dislocation.

The largest changes in GMFM scores between pre- and five years postoperatively were seen in children in GMFCS levels I-II and the least changes were seen in children in GMFCS levels IV-V. In GMFM-66 scores children in the GMFCS levels I-II, III and IV-V showed changes over the five years ($p<0.001$).

GMFM-66, GMFM-88 total and goal total scores showed significant changes using the Wilcoxon signed rank test ($p<0.001$) for the group as a whole at 1, 3 and 5 years postoperatively. There were no statistically significant differences (Wilcoxon signed ranks test) in GMFM-66 scores during the first six months, either for the whole group or for children in GMFCS I-II, III and IV-V. For children in GMFCS levels I-II, changes were not statistically significant during follow-ups. Children in GMFCS III showed significant changes after 3 and 5 years postoperatively ($p=0.005$) and for children in GMFCS IV-V at 18 months ($p=0.001$), 3 and 5 years postoperatively (both $p=0.002$).

The PEDI results for the group ($n=30$) showed statistically significant changes in the dimensions Functional Skills and Caregivers' Assistance for the domains Self-care and Mobility. Statistically significant changes ($p<0.001$) for scaled scores in both

dimensions and domains were detected between scaled scores preoperatively and all postoperative follow-ups (pre to 6 months, pre to 12 months, pre to 18 months, pre to 3 years and pre to 5 years postoperatively) for the whole group and for children in GMFCS IV-V. For children in GMFCS I-II (n=8) and III (n=7), there were no statistically significant improvements.

No major complications occurred peri- or postoperatively. All children could be discharged from the hospital and regional habilitation unit after 3½ weeks according to the preoperative plan. No new urinary tract problems, including incontinence, were present after the SDR-operation. Many children had problems with constipation preoperatively, in five children the problems disappeared postoperatively. Nine still needed medication for constipation five years postoperatively.

At five years postoperatively, the two children with preoperative overweight were still obese and two others had become overweight. One child had acquired severe thinness. Three of the 11 children who were underweighted before the operation, were still underweighted.

Sensory problems ascribed to the surgical procedure were dys- or hyperaesthesia, which had disappeared, in all but three children, six months postoperatively. One child had recurrent hyperaesthesia and flexor spasms during febrile infections.

Orthopaedic surgery after selective dorsal rhizotomy (Paper II)

Bilateral adductor tenotomy was performed on one child two years before SDR. No other orthopaedic operations were performed before SDR. At the five-year follow-up, 15 children (42%) had undergone 42 orthopaedic surgical interventions in the lower limbs, 12 were operated once (1.1-5.0 years after SDR) and three children had surgery on two occasions. The most common surgical procedures were subtalar arthrodesis 17 of 42 (41%), achilles tendon lengthening 12 of 42 (29%) and adductor-psoas tendon lengthening 4 of 42(10%). Foot-ankle surgery accounted for 33 of the 42 (79%) interventions and surgery addressing the structures around the hip accounted for 7 of the 42 (17%) interventions. No spinal surgery had been performed after the SDR.

Ten of the 20 children undergoing SDR between 1993 and 1995 with a mean age at SDR of 4.7 years, had undergone orthopaedic surgery, compared with five of the 15 children operated on between 1996 and 1999 with a mean age at SDR of 4.3 years. Mean age at SDR was 4.9 years in the 15 children undergoing orthopaedic surgery, and 4.0 years in the 20 children without orthopaedic surgery. Of the 15 children who underwent orthopaedic surgery, seven were in GMFCS level II, five were in GMFCS III and three were in GMFCS IV. Children with walking capacity mostly had foot surgery. Subtalar arthodesis and/or achilles tendon lengthening was noted in six out of seven children in GMFCS II. Children without walking capacity mostly had hip surgery, mainly adductor-psoas tenotomy.

Longitudinal construct validity of the GMFM-88 total score, goal total score and the GMFM-66 in a 5-year follow-up (Paper III)

At six months postoperatively ES and SRM were small (≤ 0.5) for all of the three GMFM scoring options, for children in GMFCS levels I-III and IV-V and for the group as a whole. At 12 months, children in GMFCS level I-III assessed with GMFM-88 total score showed large changes (ES 0.8 and SRM 1.3) as well as GMFM-88 goal total scores (ES 0.9 and SRM 1.2). Less change was seen with GMFM-66 at 12 months (ES 0.3 and SRM 0.8). For children in GMFCS levels IV-V, both GMFM-88 total and goal total score showed large changes at 12 months postoperatively (ES 0.8 and SRM 0.9) and GMFM-66 showed less change (ES 0.4 and SRM 0.7) (Paper III, Table 3).

At 18 months, children in GMFCS I-III assessed with GMFM-88 total and goal total score showed large changes (ES 0.8 and SRM 1.1, ES 0.8 and SRM 0.9 respectively). GMFM-66 showed less change (ES 0.5 and SRM 0.8). Children in GMFCS IV-V assessed with GMFM total and goal total scores, showed large changes (ES 1.0 and SRM 1.1, ES 1.1 and SRM 1.2 respectively) while ES for GMFM-66 showed less change (0.6) and SRM large change (1.0) (Paper III, Table 3).

At three and five years postoperatively, all three GMFM scoring options showed large changes for both GMFCS I-III (ES range 1.0-1.6 and SRM range 1.0-1.2) and GMFCS IV-V (ES range 1.0-1.6 and SRM range 1.0- 1.7) (Paper III, Table 3).

Gait Profile Score and Movement Assessment Profile in a longitudinal follow-up in children with cerebral palsy (Paper IV)

The GPS displayed less deviation in gait pattern after five years compared to preoperatively in six out of seven children. The changes in GPS in four children were small during the first year (Figure 5).

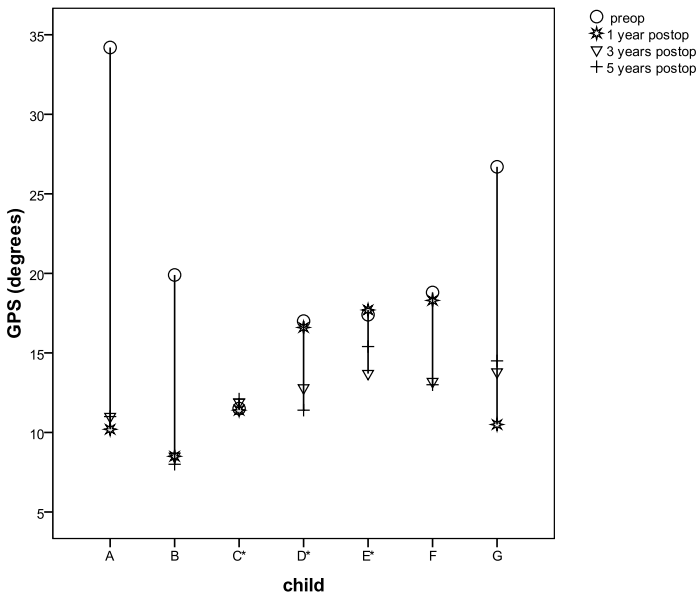

Figure 5.
Mean Gait Profile Score (GPS) for Three-dimensional gait analysis data of each individual child at preoperative measurement and follow-up at 1 year, 3 and 5 years postoperatively. Preoperatively (preop), postoperatively (postop), preoperative Botulinum toxin A injections (*).

Child A (Figure 5) was selected as a case to illustrate development of MAP over the five years. In the preoperative MAP, left foot progression was more deviated than the right (Paper IV, Figure 2). At one year, the MAP identified less deviation in ankle dorsiflexion and foot progression. Also the deviation in hip abduction was increased compared to preoperative. At three years postoperatively an increased deviation of the foot progression compared to one year postoperatively was seen whilst other improvements were maintained. Preoperatively, the GPS showed a large deviation mostly originating from the ankle and foot. Less deviation than preoperatively shown by the GPS was seen at the follow-ups.

Functional outcomes 10 years after selective dorsal rhizotomy (Paper V)

The median muscle tone was reduced for hip adductors, hamstrings, hip- and plantarflexors between pre- and 10 years postoperatively. The changes within the group in muscle tone depended on preoperative levels of tone for hip ($p<0.001$). The higher the muscle tone preoperatively, the larger the decrease was seen at 10 years postoperatively.

Changes in PROM were small between pre- and 10 years postoperatively in hip abduction, popliteal angle, knee extension, and dorsiflexion of the foot for the group. The changes within the group depended on the preoperative joint angles ($p<0.001$). Children with the largest PROM preoperatively decreased their PROM the most (Paper V, Table 3).

Changes within the group in PROM of popliteal angle, knee extension and dorsiflexion of the foot depended on GMFCS levels ($p=0.02$, $p=0.05$ and $p=0.05$ respectively) (Paper V, Table 3). For PROM in knee extension and popliteal angle, a higher GMFCS level (more functional limitations) lead to larger changes (decrease). However, with respect to dorsiflexion of the foot, the higher the GMFCS levels, the smaller changes (decrease) in PROM. Dorsiflexion in the foot was mainly maintained for the group as whole.

The mean GMFM-66 score increased between pre- and 10 years postoperatively (Paper V, Figure 4). Changes in GMFM-66 score within the group depended on preoperative GMFCS level ($p=0.008$), preoperative GMFM-66 score ($p= 0.045$) and age at SDR ($p=0.017$). The higher the GMFCS level and age at SDR, the less change in GMFM-66 was seen at 10 years post- compared to preoperative value. The higher the preoperative GMFM-66 scores, the larger changes were seen in GMFM-66 scores at 10 years postoperatively.

The mean development of GMFM-66 for children in GMFCS levels II, III and IV undergoing SDR showed a higher score for GMFCS levels II and IV at older ages compared to the children in the OMG group. Children undergoing SDR in GMFCS level III had a similar GMFM-66 score as the children of the OMG group at older ages (Figure 6).

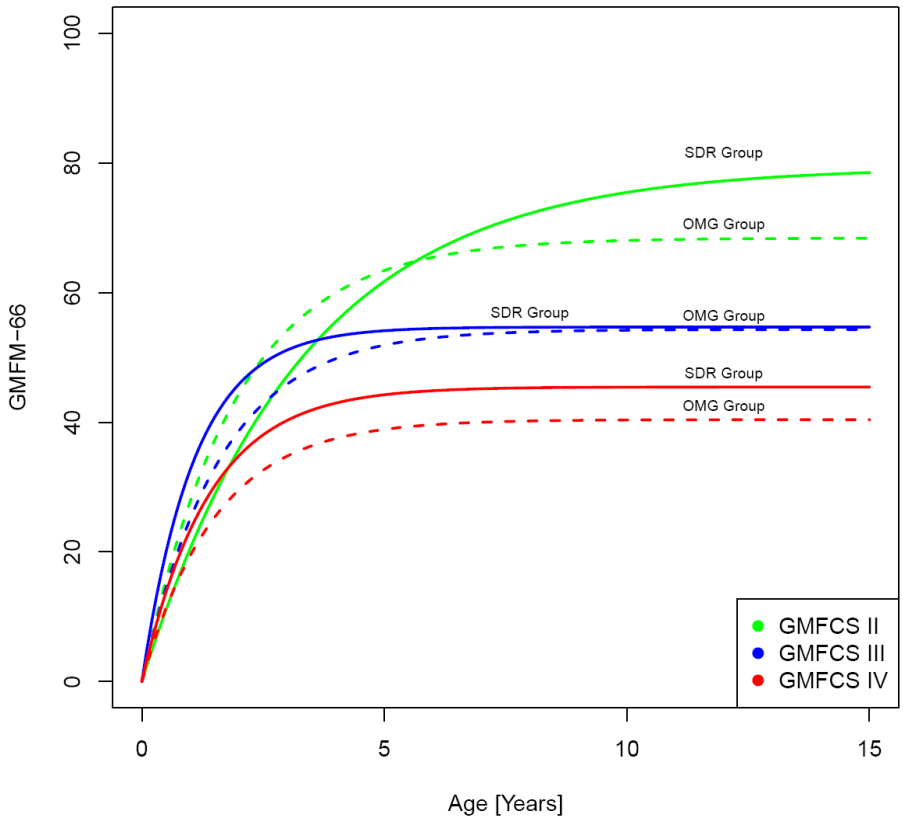

Figure 6.
Mean development of Gross Motor Function Measure (GMFM-66) as a function of age for Gross Motor Function Classification System (GMFCS) levels II (n=7), III (n=8) and IV (n=12) of the SDR group compared to the predicted mean development of GMFCS II (n= 80), III (n=122) and IV (n=137) for the Ontario Motor Growth (OMG) group. Reference values from Hanna et al. [113] are used with permission of John Wiley and Sons to produce reference curves of gross motor development of the SDR group.

Preoperatively 11 children used manual wheelchairs or crawled/rolled, 12 walked with walkers and six walked independently. At 10 years postoperatively, six adolescents used wheelchairs only, five walked with walkers shorter distances and used wheelchair longer distances, eight walked with walkers, sticks or crutches and 10 walked independently. A larger diversity of mobility devices were seen at the 10 year follow-up compared to preoperatively.

The FMS showed that all 10 adolescents in GMFCS-E&R I-II walked independently on all surfaces at home and at school. Eight of them walked independently in the

community, whilst two chose to use a wheelchair. Children in GMFCS-E&R III showed the largest diversity in walking aids. Two walked independently on level surfaces at home where the others used crutches or a walker. For mobility at school and for mobility in the community, one and two adolescents respectively chose wheelchairs for mobility, whilst the others walked with walkers or sticks. Three adolescents in GMFCS-E&R IV -V walked with walkers at home but chose mobility by wheelchair at school and in the community. The other six adolescents used wheelchairs for all functional mobility at home, in school and in the community (Paper V, Figures 5 A-C).

Discussion

These are long-term results from a group of children with spastic diplegia undergoing SDR combined with physiotherapy, followed during 10 years using standardized measures at specified time intervals. The standardized measures were chosen to evaluate different aspects of function and to improve understanding of the postoperative course of motor development. Outcome analyses in this thesis are mostly based on group level.

SDR results after 5 and 10 years (Papers I, II and V)

Results from Papers I, II and V show that SDR in combination with physiotherapy is a safe, effective and durable spasticity reducing treatment for children with spastic diplegia irrespective of functional limitations.

Selection criteria as suggested by Peacock & Staudt have been unchanged since the start in 1993 [98]. However, in this thesis, children with cognitive disabilities were not excluded if they had the drive to move and interact in playful treatment situations. The spasticity team at the specialist clinic is carefully selecting children for SDR based on the criteria as per Peacock & Staudt, personal and environmental factors for the child as well as experiences from children previously operated on. Continuity in the spasticity team at the specialist clinic, together with long-term perspectives of functional outcomes of the team members' intervention are considered to be of great importance. As the intervention is suitable for only a small number of children, the selection, operation and postoperative follow-up should be performed at a few centers as present. Children and families may have to travel long distances as well as physiotherapists from the local habilitation centers for the intervention, but the experience from a larger number of children can be gathered within the specialist team.

The appropriate selection criteria have been suggested to be the strongest predictor of outcome after SDR [152]. It is important to firmly establish both realistic expectations and the intervention's long-term goals with parents and children. The local habilitation team must to be involved during the whole process from selection to long-term follow-ups. It is a long-term mutual commitment and responsibility between the spasticity team at the specialist clinic and local habilitation team to provide an optimal rehabilitation lasting for several years. Most specifically, physiotherapy resources for intensified treatment and collaboration with specialist team need to be provided from a long-term perspective, at a cost to be established by the officials deciding over the long-term physiotherapy resources at the local habilitation centers.

In the current Papers, muscle tone was found to be immediately reduced after the operation and was still reduced after 10 years. Previously muscle tone was reported to be reduced after five years in accordance with results in Paper I [65, 153] and in a follow-up at a mean of 7 years.[154]. In Paper V, the amount of muscle tone reduction was shown to depend on the level of muscle tone prior to SDR. The higher the muscle tone preoperatively, the larger the reduction after 10 years. Muscle reflexes were also immediately reduced and maintained during the first five years. No other studies have shown results of maintained spasticity reduction during 10 years.

As the selection criteria excluded children with severe contractures no children had manifest contractures preoperatively. Gul et al. showed improved PROM in hip abduction, popliteal angle and knee extension at five years compared to preoperative values in accordance with results in Paper I [153]. The mean PROM of the hip, knee and foot were improved between pre- and five years postoperatively. However, a tendency of decrease in mean PROM of the popliteal angle was seen between three and five years postoperatively, especially for children in GMFCS III. At the 10-year follow-up, the mean PROMs were only slightly different to the preoperative PROM angles, indicating a decrease in PROM between five and 10 years postoperatively. The decrease in PROM in this time interval is in concordance with the natural development of PROM described in a total population of children with CP, the decreasing joint mobility with age in this study can be expected [38].

The spasticity reduction obtained by SDR combined with physiotherapy treatment in this group of children had a positive impact on PROM during the first five years after SDR. The PROM reductions between five and 10 years may have several causes. It has been shown that PROM decreases with increased GMFCS level, and as a large proportion of the adolescents were GMFCS III-V the mean PROMs were highly influenced by their values [155]. All children undergoing SDR were recommended to use a standing shell 1-2 hours daily in an optimal standing position for prevention of joint deformities and contractures. During the first five postoperative years this recommendation was followed in a majority of the children. Ten years after SDR, very few had continued to use the standing shell or other standing devices as a daily routine, which may have contributed to the development of decreased PROM. The adolescents still using different standing devices were in GMFCS levels III-V. The height growth spurt during puberty as well as the reduced focus on contracture prevention between five and 10 years postoperatively, probably contributed.

All children were also postoperatively recommended to use orthoses to stabilize the foot and ankle. The majority of the children had been using orthoses to reduce the equinus prior to SDR. However, postoperatively the conditions were changed; the load was shifted towards the medial part of the foot and the heels were in contact with the floor. The orthoses main purpose directly postoperatively to optimize the base of support in order to facilitate knee- and hipextension in the standing position.

After advice from the physiotherapist at the specialist clinic, the child's local orthotist together with the local habilitation physiotherapist decided which type to use for each individual child. Half of the adolescent still used orthoses for 10 years after SDR due to muscle weakness and imbalance of the muscles of the foot and ankle.

Measurement errors in goniometry in the lower limbs in children with spastic diplegia has been shown to be approximately five to 15 °, and with wide error margins [99, 102, 121]. During the time between measurements, changes in PROM may occur due to e.g. natural growth, stretching program, and the use of orthoses or serial casting, etc. Changes might actually be equal to the measurement errors. Even if the same physiotherapists performed the measurements over time, measurement error may be a part of the changes.

The amount of orthopaedic surgery at five years after SDR in Paper II was similar to results presented by Arens et al. with a rate of 48 % after a mean follow-up of seven years [154], but lower than Caroll et al. with a rate of 65% [76]. There are studies suggesting that age at SDR influenced the frequency of orthopaedic surgery after SDR, as the reduced spasticity at earlier ages would cause less contractures and joint deformities. Children undergoing SDR at younger ages needed ortopaedic surgery to a lesser extent [76, 77, 79]. The group of children in Paper II was small and had a shorter range of age than in these studies. However, there was a tendency to more surgery in children SDR operated at an older age.

The most frequent surgical intervention was subtalar arthodesis, representing 40% of all operations. This was similar to findings from Carroll et al. where 39% had foot stabilizing surgery [76]. Corresponding frequencies in other studies were 10% [78] and 22% [77]. The rates of hip surgery in Paper II were lower in comparison to the rates reported by Caroll et al. [76] Different rates of orthopaedic surgery between the studies may depend on different indications for surgery between centers and different patient selection criteria for SDR. As CPUP was introduced during these years more effort was put on preventive and non-invasive treatment options in order to reduce hip subluxation. Derotational osteotomy of the femur was performed in one child; four other children with increased subluxation had adductor-psoas tendon releases to prevent further subluxation.

In the five year follow-up, no children had received surgical treatment for spinal deformities. Others have found that spinal deformities were less common and/or less prominent in children undergoing SDR at younger ages than in older ages [55, 83]. Langerak et al. presented data from 30 patients who had undergone SDR 17-26 years ago and found no significant major spinal deformities [85]. On the other hand, increased frequencies of spinal deformities after SDR have been reported, suggesting specific follow-up of the spine for this group of children [81-84]. As knowledge of the natural development of spinal deformities in spastic CP is limited, and due to the conflicting results, continued monitoring of spinal deformities is needed.

Functional improvements were shown with the GMFM- 88 total and goal total score after five years and with the GMFM-66 after both five and 10 years. As expected, the largest changes in GMFM-66 were seen in children in GMFCS I-II and the least in GMFCS IV-V. This is in accordance with the motor growth curves, where in a population of all CP subtypes, development of GMFM-66 scores have been shown to depend on age and GMFCS level [112].

Motor growth curves were created for the GMFM-66 data for children with preoperative GMFCS levels II, III and IV (Figure 6). The slopes of the curves from the SDR group differ from the curves of the OMG group. Children undergoing SDR seem to have similar or better mean GMFM-66 development in older ages than children in the OMG group. The decline model suggested by Hanna et al. in children in GMFCS III-V (Figure 3) did not fit with the GMFM-66 development in the children undergoing SDR [113]. The differences might be a treatment effect, but this result must be interpreted with great caution due to the small number of children undergoing SDR in each GMFCS level. However, no other studies were found for comparison of the development of gross motor function.

PEDI was used in Paper I, to broaden the perspective of function and highlight performance; assessing what the individual child actually does in meaningful daily activities in contrast to capacity; what they can do in specific test situations which were measured with GMFM. Traditionally, improved walking has been a main goal for children undergoing SDR. However, in Paper I, the children with more functional limitations (GMFCS IV-V) never had walking as a goal. It has been shown that there is a great variability in mobility in children with CP, even within the same GMFCS level, due to contextual, environmental and personal factors [52].

The advantages of using PEDI are to explore the parent's view of the most common performance of the child in the home environment and the amount of caregiver assistance. In addition, it is a valuable tool in the rehabilitation process to define functionally realistic goals and modifications needed for independence in activities in daily life. In Paper I, the largest functional improvements appeared in the early postoperative follow- up. The improvements continued during the five years, which is in accordance with Mittal et al. [70].

The FMS also provided information on performance in functional mobility in different contexts at the 10-year follow-up. The adolescents were found to use mobility methods which require the more motor control at home, and the method requiring less motor control at school and in the community. This has previously been shown by Tieman et al. and in Paper V this was seen in adolescents with GMFCS-E&R III and GMFCS-E&R IV-V [156]. Two adolescents in GMFCS-E&R I-II chose wheelchair for mobility in the community while walking independently on all surfaces for shorter distances at home and at school. A few of the children in GMFCS-E&R III chose wheelchair for mobility in the community even if

they were able to use walker at home or in school. The use of wheelchair for mobility long distances saves energy which may be better used during other activities than walking, being able to keep up with peers, facilitates participation and being able to live a more independent life. The achievement of independent walking during childhood would not be the ideal goal for all children with CP. A more independent oriented approach has thus been suggested by Bottos et al. for interventions in earlier years by using a combination of walking aids and wheelchairs even for children with walking ability [157]. In a qualitative study by Palisano et al. the adolescents expressed that mobility was important for their self-sufficiency and their personal choices of mobility were based efficacy and safety [158]. Mobility preferences depended on environmental factors as well as personal choices, also seen in the results from the adolescents with FMS in Paper V.

It is crucial to use appropriate evaluative measures when appraising changes in function. The long-term goal of the intervention should decide which outcome measure to be used. The use of multiple measures complementing each other, covering both the ICF-CY components for a broad understanding of the effects is desirable.

In Paper I, treatment effects in different outcome measures were evaluated using non-parametric methods. Significant changes were obtained by the group as a whole but when subdivided into smaller groups, changes were not statistically significant and may have depended on type II errors. A larger group of children would be needed to examine differences between GMFCS groups using the non-parametric methods used. In Paper V, an even smaller group of children was available but linear regression analysis was performed to analyze changes in outcomes and thus the size of the groups was not as important.

Due to the small number of children and also when using non-parametric statistics it would have been more appropriate to use median values in Paper I.

Longitudinal construct validity of the GMFM (Paper III)

Gross motor function has been shown to depend on age and GMFCS levels, where the most rapid changes occur during the first four years of life and levelling off at the age of 6-7 years depending on GMFCS levels [112]. The mean age at SDR was 4.4 years and most of the children were likely to improve in gross motor function another 1-2 years before reaching their probable maximum scores at between 6 to 7 years of age. Changes in all GMFM scores were expected, due to the natural development of gross motor function and the effects of spasticity reduction in combination with physiotherapy. The expected changes in gross motor function were found appropriate when studying longitudinal construct validity in GMFM.

Twenty-two items in the lower difficulty scale from the original 88- item version were removed to create the GMFM-66. Children in GMFCS III-V who mainly perform

activities independently in items of the lower difficulty scale have fewer items to be tested in when using the GMFM-66. The GMFM-66 indicated similar longitudinal construct validity for GMFCS I-III and IV-V, suggesting that changes were equally detectable for children with less functional limitations and for children with larger functional limitations. The Rasch-analysis resulted in a more even distribution over the difficulty scale and changes in scores would be equal over the scale, as seen in Paper III. The GMFM-88 version includes more items in clinically relevant positions for children in GMFCS IV-V and the gross motor function is thus more appropriate described for them than by the GMFM-66.

The GMFM-88 goal total score has seldom been used in research reports. It was found useful in clinical practice for monitoring gross motor function changes in the clinically relevant dimensions. By identifying goal areas and deciding which dimensions to include in the goal total score, children, families and clinicians can discuss the long-term expectations of the intervention. Specific items from these dimensions can be used for short-term goal setting during rehabilitation. By using both functional and relevant goals, set together with child and family, motivation increases, as the goals are apparent and clear. The goal directed approach has been shown to be effective in physiotherapy treatment [159, 160].

By using the clinically important dimensions only, as in the GMFM-88 goal total total score, the instrument can more effectively measure the items where the child cooperates during the test and thus give a more correct picture of gross motor function. In GMFM-88 total score, all dimensions and items must be scored to give a correct estimation of gross motor function. Even if a child is able to perform, but refuses during the session, or if the physiotherapist forgets to test the item, a score of zero must be given. It is more likely that, despite being able to, a child who is able to perform difficult items is reluctant to perform items in lying and rolling- although he/she is able to. This may be the reason why the goal total score has somewhat higher longitudinal construct validity than the total score, especially at 3 and 5 years postoperatively. In the GMFM-66, not all items need be tested to obtain a representative estimation of the child's gross motor function. The software, GMAE deals with the items that are tested, and calculates the score from them. The GMFM-66 is thus even less sensitive to missing items compared to the GMFM-88 goal total score where all items in the selected dimension must be tested for a representative score.

ES and SRM are the two most commonly used methods to examine longitudinal construct validity [161]. The large SD of the scores before surgery and the small changes after 6 months resulted in low ES and SRM compared to later follow-ups. The change score and SD of the change scores (when calculating SRM) were more homogenous than the change scores and SD of the preoperative score (for calculation of the ES). This was reflected by the relatively larger SRM than ES, at least in follow-

up intervals up to 18 months after SDR. The small differences indicate that the group scores studied were similarly distributed at baseline and at follow-ups.

GPS and MAP (Paper IV)

The GPS detected longitudinal changes for the seven children followed during five years after SDR. Three children had received btx injections in the plantarflexors flexors of the foot within six months prior to SDR, to prevent further PROM restrictions while waiting for SDR. For these children, none or very small changes were seen in GPS during the first postoperative year. The temporary spasticity reduction obtained by btx was continued by the permanent spasticity reduction obtained by SDR. This is in concordance with the clinical picture where these three children walked with full/almost full foot contact at the preoperative assessment as well as in later follow-ups.

In longitudinal studies of gait in children with CP deterioration over time in joint excursions and temporo-spatial parameters have been shown [47-50]. Seven children had improved or maintained their GPS at five years compared to preoperatively. This is a better outcome than expected regarding previous studies on longitudinal development of gait in children with spastic diplegia, and may be a result of the permanent spasticity reduction by SDR combined with physiotherapy treatment. Additional treatments included preoperative btx in, postoperative serial casting and orthopaedic surgery.

The MAP was used to illustrate the longitudinal changes in one of the children. This child mainly showed changes of the ankle dorsiflexion and foot progression, especially between the measurements before and again one year after SDR. This child had no other surgical interventions except SDR, so the immediate spasticity reduction might be a result of the operation.

Since the children were young at their first 3DGA, their gait pattern varied between sessions. In 3DGA, parameters measured during gait are influenced by intra-subject variability as well as errors related to the methodology. The gait laboratory has used the same marker protocol during this period and the members of the staff are regularly trained in the placing of markers to reduce measurement errors. To decide when significant changes occur, measurement errors mentioned above must be taken into consideration as well as the variability of gait pattern. All children walked with less variability between trials as they grew older. It is likely that all 3DGA related results are more reliable in older children.

The GPS and MAP were found to be relative simple and useful tools in identifying longitudinal changes in gait patterns. However, the scores and profiles need to be used together with other measures to obtain a more complete picture of the multifaceted development of walking ability as possible. In Paper IV, the GMFM-66 and temporo-spatial parameters were used as complementary instruments.

The GPS and the MAP may be easier to understand than a selection of univariate variables when communicating results to health professionals unfamiliar with the terminology. They may also be useful when describing the deviation and development of gait patterns for children and their parents.

Methodological aspects

Randomized controlled trials (RCT), have been considered as the criteria in the field of health science as they help to isolate the effect of a single intervention. However, trials when controlling for various variables may not reflect what happens in the real world and may even overestimate the effect of treatment, especially when performed in complex multidisciplinary management of children with CP [162]. The Clinical Practice Improvement (CPI) research methodology has been applied as an alternative approach by Horn et al. [163]. It is an observational study design comprising a thorough review of care management processes including key patient characteristics as well as all treatment and care processes and outcomes. The CPI approach fits in with the Papers I, II and V, and has been found to be valuable in these types of clinical studies. It has been recommended in e.g. research of musculoskeletal issues in developmental disabilities to complement the research obtained by RCT [162].

The distinctions between GMFCS levels are clinically relevant and five subgroups would be optimal, however subgroups according to GMFCS levels were created due to the small number of children in each GMFCS level. In Paper I, the three groups GMFCS I-II, III and IV-V were used. Children in GMFCS III differ from children in GMFCS I-II in respect of e.g. development of PROM, and from children in GMFCS IV-V by the ability to walk with a walking aid. However, the number of children in each group was probably too small to detect changes especially for children in GMFCS I-II and III, and the risk of type II error was high. When the results from the children in GMFCS I-II were merged with GMFCS III into one group changes were statistically significant. Different statistical methods were used in Paper V, which allowed for analysis according to the five GMFCS levels.

In Paper III, the two groups consisted of children in GMFCS I-III and IV-V so as to obtain two groups of similar sizes. Children in GMFCS III were included with children in GMFCS levels I and II in as they all walk with or without walking aids in contrast to children in GMFCS IV-V who mainly relied on wheelchair for functional mobility.

In the literature, SDR is recommended to be combined with intensified physiotherapy to improve functional outcomes when spasticity is reduced. In the meta- analysis of the three RCTs, children undergoing SDR received 2-3 hours of individualized physiotherapy per week during the first postoperative year [73]. However, how the rest of the hours of the week are spent is likely most important. Today, other approaches for physiotherapy interventions are available and in practice.

By the family centred approach in child health services, the child and family are setting the goals together with the physiotherapist. Functional goals for therapy interventions can be identified. The goals are relevant for the child and family and thus make sense and are applicable in the daily environment of the child, involving persons around the child in daily life. By identifying clinical goals within daily activities, more repetitions can be performed in relevant environments and situations during the day for the child than when treatment is performed once or twice a week. It has been shown that goal directed functional physiotherapy in children with CP has been successful [159, 160, 164].

For the children undergoing SDR in the thesis, individually selected functional goals have been set for long- and short-term with child, family and local habilitation physiotherapist. In addition leisure activities and an active lifestyle has been encouraged. The local habilitation physiotherapist has had an important role tailoring treatment interventions into the child's environment in daily life situations. The specialist physiotherapist contributed with specific knowledge in postoperative SDR physiotherapy, with experience from previous children, as well as recommendations on orthoses and aids. Intensified physiotherapy, lasting about 2 years, has been recommended. However, the increased amount of treatment provided by the habilitation physiotherapist has not been examined. During the first two year the local habilitation physiotherapists have been most active in the individual treatment. Gradually, personal assistants or extra resources at day care or school has performed exercises tutored by the habilitation physiotherapists. At the 10-year follow-up, the majority of the adolescents had no regular individual physiotherapy treatment sessions, except those few adolescents attending special schools. Adolescents participated in physical education at school, disability sports and/or exercised at gyms.

Selection of outcome measures

In longitudinal studies, the same measures should be used, despite of the introduction of new and perhaps better instruments over the years.

Assessment of muscle tone according to the Modified Ashworth scale modified by Peacock & Staudt, was chosen in order to be able to compare with their results. The psychometric properties of the scale have not been studied. Reliability of the similar Ashworth scale, as modified by Bohannon and Smith, has been found to be low to moderate in children with CP [99, 103]. The modified Tardieu scale has recently been found to better separate spasticity from movement restrictions within the muscular structures [99-101] and also better reliability than the Modified Ashworth by Bohannon and Smith was shown [99]. Today the modified Tardieu scale would probably be a better tool for assessing muscle tone in children undergoing SDR.

The selective voluntary motor control (SVMC) has been clinically examined in the children undergoing SDR, but valid and reliable quantification has not previously

been available. However, the Selective Control Assessment of the Lower Extremity (SCALE) assessment of SVMC has recently been shown to be valid and reliable in children with CP [106]. The tool is thus highly relevant for children undergoing SDR and will most likely be involved in the test battery for pre -and postoperative functional outcomes.

In 1993, the GMFM-88 was newly introduced and the total and goal total scores were used. Since then, the GMFM has been undergoing further development and the GMFM-66 became available in 2002 [111]. By using the GMFM-88 assessment and the software GMAE, GMFM-66 scores were easily obtained. All GMFM-88 assessments can be transformed into GMFM-66, even if this was not possible when the actual assessments were made.

The GMFCS was first available in 1997. It was considered possible to retrospectively classify the performance of gross motor function by reviewing medical records and video recordings from the preoperative assessment.

In recent years there has been an increased interest in muscle strength in children with CP and several studies report improved function after strengthening programs [30-32, 165]. Measurements of strength are commonly performed by a handheld dynamometer. However, reliability has been shown to be poor in two recent studies of children with CP between the ages 5.5-14 and 5-17 years of age [166, 167]. For children intended for SDR, assessments of underlying strength are performed preoperatively by functional assessments, e.g. count of repeated sit to stand, also items in GMFM (e.g. sit to stand, walk up and down stairs, one-leg standing) are used to assess functional strength, however no standardized measures were included in the test battery.

Muscle strength has been shown to correlate to GMFM scores [30-32, 69, 165, 168]. Improvement in GMFM scores in Papers I and V might be interpreted as improvements due to improved strength. Even if selected GMFM items may improve due to increased strength, the GMFM was not designed, and is not a valid instrument, for evaluating strength.

Children undergoing SDR are very young and with available techniques for measuring strength, such as handheld dynamometer and biodex, the child needs to cooperate in order to obtain reliable results. Valid and reliable instrumented tools for measuring strength in very young children with CP are needed.

In order to assess functional goals set together with child and family there may not be standardized instruments for evaluation. Families rarely state their goals in terms of impairment; rather most often they are concerned with promoting activity or participation to enhance their lives. The Goal Attainment Scaling (GAS)[169] and Canadian Occupational Performance Measure (COPM)[170] have been shown to detect clinically relevant changes in activity and participation for children with CP

[171]. The GAS and COPM may be useful in the short-term goal setting for children undergoing SDR but has until now, not been used.

Clinical implications

The spasticity team at the specialist clinic has experience in selecting children suitable for SDR. Experience is based on structured regular longitudinal follow-up with standardized outcome measures and the same professionals participating in the team. Cooperation for a long-term optimal rehabilitation is of great importance, with a shared responsibility for follow-up and treatment between the local habilitation and spasticity team at the specialist clinic.

Limitations in PROM also need to be targeted with follow-up and active interventions between five and10 years after SDR. Only half of the children in the thesis had birth years or places of residence included in the prevention program, CPUP. The program was not fully implemented for many of the children undergoing SDR as it was recently introduced. CPUP has had great impact over the last decade by increasing the knowledge of prevention of musculoskeletal deformities in CP. Limitations in PROM in children undergoing SDR should also be early identified, as well as for all children with CP.

Functional goals prior to SDR should decide primary evaluative measures of functional outcomes. Multiple measures of body functions, body structures, activities and participation are needed. Measures of capacity and performance should be used as complementary.

GMFM-66 identified changes in children in GMFCS I-III as well as changes in children in GMFCS IV-V and may be used irrespective of GMFCS levels. However, when describing the gross motor function in children in GMFCS levels IV-V the GMFM-88 scoring options have 22 more and clinically relevant items in lying, sitting and kneeling. In children in GMFCS IV-V the choice of GMFM version should be done depending on the purpose of the measurement.

By using GPS and MAP results originating from 3DGA, the most important deviations from a typically developing group of children can be identified and explained. It may be especially useful for comparison where multiple assessments are compared over time.

Future studies

To continuously follow the group of children and adolescents undergoing SDR over the life span, would provide information of the long-term effects of SDR in combination with physiotherapy. The outcome in adult age will be a result of many contributing factors of which SDR is one. Children, adolescents and adults with CP and their families are all different and live their lives according to their own choices, which also may affect functional outcomes.

The influence of personal and environmental factors in children with CP and their families undergoing SDR may be further examined. Qualitative methodology would probably be the most appropriate for these issues

To measure functional outcomes in children undergoing SDR, valid and reliable measures covering a broad perspective of functioning are needed. The ICF-CY may be useful to provide a model to identify the component level of outcome measures. As children with spastic diplegia are a heterogeneous group with large differences in functional limitations, the choices of outcome measures can be related to the different GMFCS levels and different test batteries may be created.

The ICF-CY component participation may be further elaborated and explored for this group of children as well as health related quality of life and presence of pain.

Inter- and intra-rater reliability of muscle tone assessed according to the Ashworth scale as modified by Peacock & Staudt has not been published. A comparison with the modified Tardieu scale would be of interest as it has been shown to be more reliable for rating passive resistance to lengthening of a muscle in children with CP, even if it also has its limitations.

General conclusions

The overall aim was fulfilled by following general conclusions:

- SDR is a safe and effective method to reduce spasticity, without major negative side effects.

- SDR combined with physiotherapy provide lasting functional benefits during 5 years for children with spastic diplegia.

- At five years after SDR, less than half of the children had been undergoing orthopaedic surgery. Children with walking capacity were mostly operated with stabilizing foot surgery and children without walking capacity with tenotomies of the adductor-psoas muscle tendons at five years after SDR.

- GMFM-88 total, goal total and GMFM-66 scores showed large longitudinal construct validity in a five-year follow-up.

- The GMFM-66 could identify changes in gross motor function in children in GMFCS levels I-III and IV-V.

- The Gait Profile Score and the Movement Assessment Profile was found useful for longitudinal studies in three- dimensional gait analysis with repeated sessions.

- Muscle tone was momentarily reduced, mean PROM decreased in hip and knee, but was unchanged in dorsiflexion, 10 years after SDR.

- The children showed improved capacity of gross motor function at 10 years after SDR compared to preoperatively, and changes depended on the severity of CP by GMFCS levels and age at SDR.

- Performance of functional mobility was related to the severity of CP (GMFCS-E&R levels) and contexts at 10 years after SDR.

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