Selection criteria, short and long-term effects of Selective Dorsal Rhizotomy in Children with Spasticity

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Selection criteria, short and long-term effects of Selective Dorsal Rhizotomy in Children with Spasticity

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promotoren:	prof.dr. J.G. Becher
	prof.dr. M. Steinlin
copromotoren:	dr.ir. J. Harlaar
	dr. R.J. Vermeulen

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Chapter 1

General Introduction

Cerebral Palsy

Cerebral Palsy (CP) is an umbrella term. It does not describe one single diagnosis but emphases a group of very different disorders. Various definitions of the term CP have been given in the literature [1–14]. Whereas some criteria are constantly present in all definitions of CP – namely that CP results from pathology in the developing brain, is non-progressive and leads to physical disability – the definitions otherwise considerably differ. The motor impairments in CP are well described in all reports, but it is mostly not stated that – in addition to the motor disability – the pathology in the brain also leads to various other medical conditions. The latest definition also mentions the co-morbidities that accompany the motor disturbance and describes CP as follows: "Cerebral palsy is group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. Cerebral Palsy is accompanied by disturbances of sensation, perception, cognition, communication, behaviour, by epilepsy and by secondary musculosceletal problems" [1].

CP is the most common cause of physical disability in childhood in industrialised countries. Epidemiological Registries allow to answer questions with respect to the prevalence and the clinical characteristics of CP [15]. European [16], Australian [17] and North American registries [18] document a very similar overall prevalence, which is estimated of about 2 cases per 1000 live births. Low-birth-weight children and very-low-birth-weight children are of particular risk for developing CP [19;20]. Decreased neonatal mortality is related to improved care and has also changed perceptions of viability of these infants. These changes have resulted in a rise in the absolute number of VLBW infants at risk of cerebral palsy.

The detailed mechanisms leading to brain lesions in the fetus or the newborn which later may lead to CP often remains unclear. In the last years great effort has been invested and many risk factors, that are associated with the development of CP – such as birth asphyxia, neuronal disorders, infection in utero, chorioamnionitis, vascular lesions and coagulopathy – have been defined [21–27]. How a fetus or a newborn child responds to risk factors is influenced by the genetic makeup of the child. A number of genetic polymorphisms have been associated with CP [28]. Recently a meta-analysis which assessed the relationship between alleles and genotypes and the susceptibility for CP was performed and identified a significant association between CP and polymorphisms in the interleukin-6 gene rs1800795 [29], which might indicate that the IL-6 gene plays an important role in the pathogenesis of CP – however, the detailed underlying pathophysiological process has yet to be determined.

Depending on the clinical presentation of the motor disorder, three subtypes can be distinguished in CP: Spastic CP, Dyskinetic CP and Ataxic CP [4]. Spastic CP is by far the most common subtype [16;18;30]. According to the Surveillance Group of Cerebral Palsy in Europe (SCPE) at least two of the following clinical signs should be present in spastic CP: Abnormal pattern of posture and/or movement, increased tone and pathological reflexes [4]. Spastic CP may be either bilateral or unilateral: Bilateral spastic CP is diagnosed if the limbs on both sides of the body are involved and unilateral spastic CP is diagnosed if only the limbs on one side of the body are involved.

According to its definition [1] CP is an umbrella term which describes motor abnormalities that result from a variety of malformations or lesions in the developing brain. With the aid of neuroimaging studies – especially with Magnetic Resonance Imaging (MRI) – differing patterns of cerebral lesions or malformations can be detected, which are related to the timing of the injury during the development of the brain [31–33]. As an example periventricular white matter lesions are often observed in former preterm infants, who clinically may present with bilateral spastic CP. The unique cerebrovascular anatomy and physiology of the premature baby underlies the exquisite sensitivity of white matter to the abnormal milieu of preterm extrauterine life, in particular ischemia and inflammation [34]. Spasticity in children – however – can also be the clinical manifestation of lesions or malformations in the spinal cord, a possibly treatable condition in the brain (such as hydrocephalus or vascular malformations) or a progressive disorder (such as metabolic disease or hereditary familiar spastic paraplegia). As the clinical examination does not necessarily allow to distinguish one from another, the Standards Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurology Society recommended neuroimaging studies in the diagnostic work-up of all children suffering from CP [35].

Spasticity and Spasticity Treatment

The most common cited definition of spasticity is that of Lance et al, who stated that "Spasticity is a motor disorder, characterized by a velocity-dependent increase in the tonic stretch response (muscle tone) with exaggerated tendon jerks, resulting from hyperexcitability of the stretch reflex, as one component of the upper motor neuron syndrome"[36]. Although Lances definition of spasticity has been criticized for being too narrow and describing spasticity only as a form of muscular hypertonia [37;38], it points out that spasticity is simply one single component of the upper motor neuron syndrome (UMS) and other features of the UMS, such as muscle weakness, loss of dexterity and selectivity of movement and increased fatiguability (=negative features) as well as extensor spasms, flexor spasms, postural reflexes, mirror movements and co-contractions (=positive features) often occur in conjunction with

spasticity [38]. The symptoms of the UMS are mostly due to lesions in the parapyramidal tracts. These descending pathways synapse upon interneural networks in the spinal cord that control spinal stretch reflexes as well as flexor and extensor reflexes. Abnormal processing of spinal reflexes, which results from a disturbance of supraspinal inhibitory and excitatory inputs (overactivity of excitatory inputs) and produce a state of net disinhibition of the spinal reflexes contributes to most of the positive features of the UMS syndrome – which is also the case for spasticity [38].

In patients with spastic CP different treatment modalities have been introduced to reduce spasticity [39–42]. Besides physical therapy and occupational therapy, orthotic management plays an important role. The medical management of spasticity includes focal and generalized treatments which can be reversible or irreversible. Generalized treatments include a large variety of spasmolytic drugs – such as baclofen, tizanidine, dantrolene and benzodiazepines. Spasmolytic drugs may have a poor passage through the blood-brain barrier and are often associated with side effects. Intrathecal baclofen therapy (ITB) represents a well tolerable and effectiveefficacious alternative for the management of generalized and intractable spasticity. Focal treatments include intramuscular injections of botulinum toxin, chemical neurolysis with phenol or alcohol and orthopedic surgery. Furthermore, different lesioning neurosurgical procedures such as peripheral neurotomy, radiofrequency lesioning of the dorsal root ganglion and selective dorsal rhizotomy (SDR) are used to decrease spasticity in patients with spastic CP.

In the management of children with spastic CP, the selection of the treatment modalities is complex and the treatment goal depends on various factors, such as the age of the child, cooperation of the patient, presence of comorbidities, functional limitations and accessibility of a rehabilitation setting. The treatment should not primarily focus on the reduction of the spasticity per se – but rather include well defined treatment goals, which influence different components of the patient's health condition. Furthermore it often remains unclear to what extent the spasticity itself mainly contributes to limitations in functioning and other features of the UMS (such as weakness and loss of dexterity) might have a larger influence on gross motor function than spasticity [43] and the relation between spasticity, strength, gross motor function and functional outcome is very complex [44]. Moreover, spasticity might even have a positive influence on a gross motor function – for example it might have a stabilizing function in patients with profound weakness but preserved walking or standing ability. Therefore spasticity treatments – especially when irreversible – must be indicated with caution.

Selective Dorsal Rhizotomy

Dorsal rhizotomies are neurosurgical treatments which are based on a neurosurgical interruption of the afferent input of the monosynaptic stretch reflex. Already in 1898 Sherrington et al showed, that the muscle tone could be reduced when the dorsal roots of decerebrated cats was trans-sected. The German neurosurgeon Othried Foerster performed a trans-section of the posterior roots in patients with spastic CP already in 1904 and described a decreasing muscle tone in all patients [45]. However, due to undesired effects on sensory and sphincter function the indication was very limited and the operation technique was abolished. To reduce these undesired effects, different neurosurgeons developed more selective operation techniques, in which the dorsal roots are split into separate rootlets and only one part of the rootlets are trans-sected – leaving the other part intact and therefore perceive sensory function. Gros et al. [46] and later Ouaknine [47] introduced a technique that consisted of sparing one rootlet in five of each root respectively one third of two thirds from L1 to S1. Gros [48], Privat [49] and Frerebeau [50] proposed a topographic selection of the rootlets to be trans-sectioned. However, the most widely used operative technique is the so called "selective dorsal rhizotomy" (SDR) – which was introduced by Fasano [51] and later-on modified by many other neurosurgeons [52–54]. This method is based on intra-operative bipolar electrical stimulation of the dorsal rootlets and the analysis of muscle responses on electromyography (EMG). Pathological responses – such as permanent tonic contractions or large spatial diffusions to distant muscle groups or to the contra-lateral side are considered to belong to the disinhibition of the spinal circuits and to be responsible for spasticity. The rootlets to be trans-sected are selected according to the EMG responses – and in some centers also based on the manual palpation of muscle contraction. In the VU University Medical Center SDR was introduced in 1998. The operative procedure in our center consists in exposing the dorsal roots L2–S1 and separating them into different rootlets after a laminotomy L2–L5 and opening of the dura. The trans-section of the rootlets is performed after electro-stimulation, according to palpable muscle contraction and EMG response. At most, 50% of the rootlets are transected on one level. To prevent sexual and bladder disturbances, rootlets/fascicles showing electrical response after stimulation of the penis/the clitoris are spared (see figure 1a and 1b).

Measuring Outcome after SDR

SDR has mainly been performed in children with bilateral spastic CP and earlier studies were mainly focused on the assessment of spasticity. However, later studies involved a multidisciplinary evaluation and described various part of the patients health condition. In 2001 Steinbok published a systematic review and provided an extensive overview concerning the outcome after SDR in children with spastic CP [55]. Most of the reported studies did not include a control and consisted in prospective or retrospective case series – and therefore do not allow a causative conclusion with respect to the effect of the intervention, but the review also included three randomized controlled trials (RCT's) which compared the outcomes after SDR combined with physical therapy with physical therapy alone [56–58]. It was shown that SDR has a positive effect on passive range of motion [56;57;59–67], strength [56;61;68–71], spasticity [56–59;61;62;65–79], gait performance [57;60;80–87], Gross Motor Function (assessed with the Gross Motor Function Measure [GMFM][88]) [56–58;62;78], sitting ability [66–68;71;79;87;89;90] and suprasegmental motor function [66;67;72;74;75;79;89;91–96]. It was concluded that there is strong evidence for benefits of SDR with respect to lower limb spasticity and lower limb range of motion [55]. With respect to gross motor function the review was not conclusive. Whereas two of the RCT's showed a significantly larger improvement of the GMFM in the intervention group (SDR and physical therapy) [56;57], one study did not find a difference in the functional improvement after SDR between the intervention group and the control group (physical therapy alone) [58]. In 2002 McLaughlin et al. pooled the data of the three RCT's and found that 9 months to 2 years after SDR there was a significantly larger reduction of spasticity and a significantly larger improvement of the functional abilities in the intervention groups compared to the control groups [97].

Since the publication of the systematic review published by Steinbok in 2001 [55] various articles, which describe the outcomes after SDR in children have been published. Most studies described the outcomes in children suffering from bilateral spastic CP. However, a positive effect of SDR was also documented in patients with transverse myelitis [100], neurodegenerative disease [101], stroke [102], unilateral spastic CP [103] as well as in patients with bilateral spasticity and normal MRI findings due to due to different etiologies [104]. Except for the Meta-Analysis mentioned above [97] and one RCT that compared the effect of two anesthetic drugs on the prevention of brisk hyperactive response during SDR [105] – most studies represent case reports as well as retrospective or prospective case series. Only in a few prospective case series the outcomes of patients undergoing SDR were compared to a control group who did not undergo an intervention [97;106–111] and only few studies compared the outcomes after SDR with other interventions, such as intrathecal baclofen therapy [112] or orthopedic surgery [113]. One study compared the outcomes of SDR with respectively without

electrophysiological guidance [114] and one study compared the outcomes after SDR with respectively without preoperative physical therapy [115]. The remaining studies lacked a control group. The study size varied considerably and ranged from 1 to 208 participants. Below the results will be summarized and the outcomes will be classified according the domains of the International Classification of Functioning, Disability and Health (ICF) [98] and the International Classification of Functioning for Children and Youth (ICF-CY) [99]. The ICF is a classification of health and health-related domains. These domains are classified from body, individual and societal perspectives by means of two lists: a list of body functions and structure, and a list of domains of activity and participation. Since an individual's functioning and disability occurs in a context, the ICF also includes a list of environmental factors (see figure 2 and table 1). The ICF-CY was developed to be structurally consistent with the ICF for adults, but in addition includes include developmental aspects.

Body structure & function

In agreement with the meta-analysis published by McLaughlin et al. [97] in the body structure & function domain a significant reduction of spasticity was reported by several study groups [103;109–112;114–130]. In addition an improvement of passive range of motion [112;114;115;117;119;121–123;126;128;129;131], changes in strength [107;110;111;114;117;124], improved gait performance [103;106;107;119;123;124;126;127;132–136], voluntary movements [130], decreased hip migration [121;137], improved smooth pursuit eye movements [138] and improvements of electrophysiological outcome measures [105;118;122;139] were documented.

Activity

Different outcome measures, such as the GMFM [88] and the Pediatric Evaluation of Disability Inventory (PEDI) [140] have been used to assess the effect of SDR in the activity domain and in accordance to the meta-analysis a significant improvement of functional performance after SDR was observed by various study groups [104;107;108;112–115;117;117;120–122;124;126;128;130;131;133;141;142].

Participation

Outcomes in the participation domain only were rarely reported [123;126;128;133;143]. In most studies the outcome measures that have been used were not uniquely designed to measure participation and assessing participation after SDR was not the primary goal of the study. Most studies used the PEDI [140], which mainly provides a comprehensive assessment of functional skill development but also assesses the level of independent performance of functional activities in the child's environment. Chan et al.[126] used the Canadian Occupational Performance Measure (COPM) [144]. The COPM is a tool that measures individually identified problems rated on performance score and was developed for the evaluation of participation restrictions. This study

showed improvements in social participation 6 and 12 months after SDR in a group of 20 children with spastic CP. However, no control group was evaluated and it remains unclear to what extend the improvements in participation can be attributed to SDR.

Whereas most of the outcome studies report the outcomes after SDR on a short term period, outcome data on longer term now became available and follow up studies, which reported a positive effect of SDR up to 20 years after SDR have been published recently [106;130;145]. However, various studies also reported adverse events after SDR; spinal abnormalities are often observed after SDR [145–149] and urinary complication, such as bladder incontinence may occur. Despite its widespread application in North-American, African, Asian and European countries many questions with respect to SDR still remain unsolved. For example the selection criteria vary considerably between single centres and are rarely documented in detail in the different studies. To date the selection criteria for SDR have not been studied systematically. However, as SDR represents an irreversible intervention and as spasticity may also have a stabilising function – especially in patients who are ambulatory – selecting those patients who have the largest benefit and excluding those who might sustain deterioration in function is crucial. Furthermore, although it is now known that SDR has a positive effect on spasticity and functional performance in patients with bilateral spastic CP, its role in patients suffering from spasticity due to other etiologies remains unclear. At last – despite the latest publications– the impact of SDR on long term outcomes and the causal relation of possible adverse events – such as spinal abnormalities after SDR remain unclear.

FIGURE 1a AND FIGURE 1b **Selective Dorsal Rhizotomy**

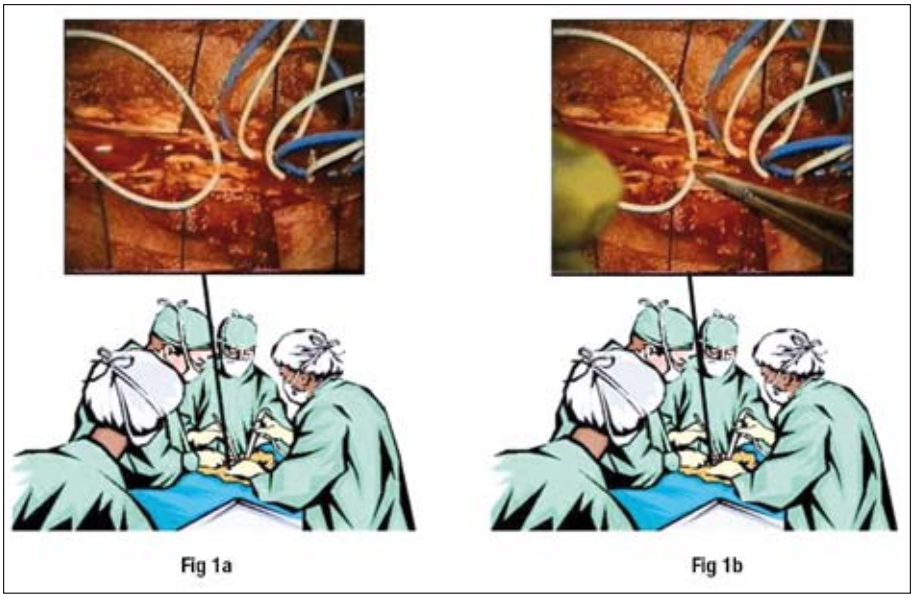
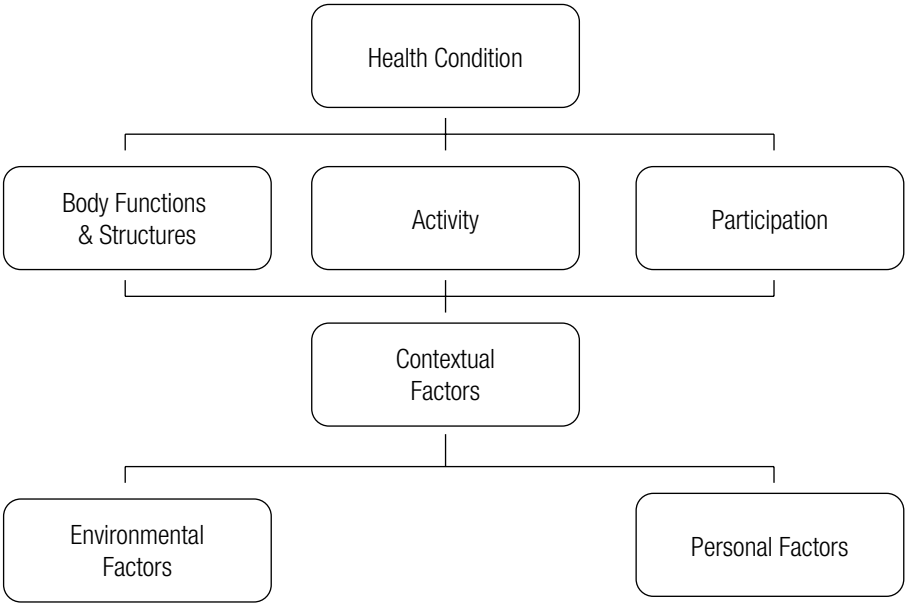


Figure 1a and 1b show the operation field of a patient during SDR. After a laminotomy and opening of the dura the dorsal roots are exposed and separated into different rootlets. The different rootlets in one anatomical level are exposed and marked with colored strings (red, blue and white) (Figure 1a). After the separating and marking the rootlets an electrostimulation is performed (not shown). Rootlets showing pathological response in EMG and/or muscle contraction are defined. The trans-section of the rootlets showing pathological responses is performed on each anatomical level (Figure 1b). At most 50% of the rootlets are transsected. Rootlets/fascicles showing electrical response after stimulation of the penis/the clitoris are spared.

TABLE 1 **Components of the International Classification of Functioning, Disability and Health (ICF) [98] and the International Classification of Functioning for Children and Youth (ICF-CY)[99]**

ICF Domain	Definition
Body Structures and Body Functions	*Body Structures* are anatomical parts of the body such as organs, limbs and their components. Impairments are problems in body structure as a result of deviation or loss. *Body Functions* are the physiologic functions of body systems. Impairments are problems in body function as a result of deviation or loss.
Activity and Participation	*Activity* is the execution of a task or action by an individual. Activity limitations are difficulties an individual may have in executing activities. *Participation* is involvement in a life situation. Participation restrictions are problems an individual may experience in involvement in life situations.
Contextual Factors	*Environmental factors* make up the physical, social and attitudinal environment in which people live and conduct their lives. They can be viewed as facilitators (positive influence) or barriers (negative influence). *Personal factors* are the particular background of an individual's life and living and compromise features of the individual that are not part of a health condition.

FIGURE 2 **Components of the International Classification of Functioning, Disability and Health (ICF) [98] and the International Classification of Functioning for Children and Youth (ICF-CY) [99]**



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Chapter 2

Aims and outline of the thesis

The present thesis was conducted to assess short and long-term outcome after SDR in children with spasticity due to different etiologies. The selection process for SDR is crucial. The decision whether to perform SDR or not is mainly based on clinical findings (such as the presence of spasticity or the absence of weakness). However, only little is known about the influence of preoperative patient characteristics (such as the preoperative level of functioning) on outcome after SDR. The effect of SDR in children who suffer from bilateral spasticity due to other etiologies than CP has not been documented in detail previously. Furthermore, neuroimaging has not been studied in detail in patients who underwent SDR and it remained unclear to what extend preoperative MRI would help to identify patients for SDR. As mentioned the short term effects of SDR has been extensively studied in children diagnosed with spastic CP and it has been shown that SDR reduces spasticity and improves gross motor function. Only recently long-term follow up studies that provide information on the effect of SDR on longer term have been published. Based on this background, the following research questions were formulated:

A. What are the effects of SDR in children who suffer from spasticity due to other etiologies than CP?
B. Can SDR represent a treatment modality in patients with progressive spasticity due to neurodegenerative disease?
C. Can neuroimaging studies help to select patients for SDR?
D. Is there a relationship between preoperative MRI findings with the change in functioning after SDR?
E. Is there a relationship between the preoperative level of functioning and the improvement of gait performance after SDR?
F. What are the long term effects and adverse events of SDR?

Study population

The study cohort included children with bilateral spasticity that underwent SDR in the VU Medical Center, Amsterdam, the Netherlands. All patients suffered from bilateral spasticity of the lower extremities. The etiology of the spasticity differed, however SDR was only performed in patients with bilateral spastic CP. The selection criteria were dependent on the goal setting. Most patients were in the GMCFS levels I to III and the main goal of SDR was to improve gait performance. In ambulatory patients SDR was only performed when A. spasticity was significantly interfering with walking performance, B. spasticity was bilateral and spasticity was present in at least six muscle groups of the legs, C. There was sufficient force in the quadriceps femoris muscle and the hip extensors, D. There were no severe structural orthopedic deformities or contractures at hip, knee, or ankle, E. moderate to good selective

motor control in the lower limbs was present and F. There was a good support from parents and rehabilitation setting. In a small subgroup of nonambulatory patients with spasticity due to neurodegenerative disease the goal of the SDR was to improve the ease of care.

Procedure

In the VU Medical Center SDR is performed since 1998. The same neurosurgeon (WO) performed the operation for all patients. After a laminotomy of L1–L5, the dura is opened to expose the dorsal roots L1–S2. Each dorsal root is separated in three to four rootlets. Consecutively, each rootlet is stimulated separately with electrostimulation, so that abnormal, exaggerated responses of the stimulated muscles can be revealed. The responses caused by electro-stimulation are recorded by EMG and by manual palpation of the muscle activity. Low threshold of muscle response and eradiation of the muscle responses are used as criterion to select rootlets for the rhizotomy. At most, 50% of the rootlets are transected on one level. To prevent sexual and bladder disturbances, rootlets/fascicles showing electrical response after stimulation of the penis/ the clitoris are spared.

Outline of the thesis

Chapter 3 aims to describe the outcomes after SDR in patients who suffer from progressive spasticity and included two case reports of patients who suffered from bilateral spasticity due to neurodegenerative disease and who were treated with SDR for the ease of care.

Chapter 4 describes the value of preoperative MRI findings in patients with bilateral spasticity and walking ability due to different etiologies.

In **chapter 5** the reliability and the validity of a custom made software program that is routinely used in the VU medical center to assess gait kinematics in patients with spastic CP before and after SDR is examined.

Chapter 6 describes the short term outcome with respect to gait after SDR.
In this chapter it is also depicted to what extend the preoperative level of functioning is related to the changes of gait characteristics after SDR.

Chapter 7 consists of a systematic review which illustrates the long-term outcomes and adverse events in children with bilateral spastic CP who underwent SDR.

Chapter 8 is the general discussion. In this chapter the findings are discussed in relation to the research questions. Furthermore the clinical implications and directions of future research are illustrated.

Chapter 9 is the summary of the thesis

Chapter 10 is the Dutch summary of the thesis

Chapter 3

Effectiveness of Selective Dorsal Rhizotomy in two Patients with Progressive Spasticity due to Neurodegenerative Disease

Sebastian Grunt, Marjo S. van der Knaap, Willem J.R. van Ouwerkerk, Rob L.M. Strijers, Jules G. Becher and R. Jeroen Vermeulen

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Abstract

Selective dorsal rhizotomy (SDR) is a neurosurgical procedure, which reduces spasticity in the legs. The effect of SDR has mainly been studied in children with spastic cerebral palsy with preserved walking ability. Little is known about the outcome of SDR in patients with neurodegenerative disorders. We report the clinical course after SDR in two patients with progressive spasticity. After SDR, leg spasticity was effectively and persistently reduced in both patients, facilitating care and improving sitting comfort. However, spasticity of the arms and other motor disturbances, such as spontaneous extension spasms and the ataxia increased gradually.

SDR leads to a reduction in leg spasticity in patients with a neurodegenerative disease. Other motor signs are not influenced by SDR and may increase due to the progressive nature of the underlying disease.

Introduction

Upper motor neuron lesions in the brain or spinal cord lead to a reduction in the inhibitory impulses on lower motor neurons, resulting in spasticity. Spasticity is defined as a velocity-dependent increase in tonic stretch response, with excessive tendon jerk reflexes[1]. It interferes with motor performance and leads to problems in patient care, sitting problems and pain. In children spasticity is most commonly observed with the context of cerebral palsy (CP). Other conditions that result in severe spasticity, such as spinal cord injury, traumatic brain injury, hereditary spastic paraparesis, and other neurodegenerative diseases are less common in children and young adults. A reduction in spasticity plays an important role in the medical management of patients that suffer from spasticity, irrespective of the cause. Different treatment modalities, such as oral anti-spastic medication, orthoses, intramuscular application of botulinum toxin A, and neurosurgical procedures such as intrathecal baclofen (ITB) and selective dorsal rhizotomy (SDR), have mainly have been studied in children with CP[2,3].

SDR is a neurosurgical procedure that reduces spasticity in the legs. By incomplete (25–75%) transsection of the (sensory) posterior lumbosacral rootlets, SDR reduces the excitatory input from the legs that enters the spinal cord. Several studies have reported a significant reduction in lower limb spasticity[4–8], improvement in functional performance[4–13], better performance with respect to the activities of daily life[6,9,12,13,14] and significant improvement in gait pattern[6,10,13,15–18] after SDR in children with CP. However, there is little knowledge about the effects of SDR in children with a progressive neurological disease.

We report the outcome of two patients with neurodegenerative disease. These case-reports are intended to illustrate the possibility of SDR as treatment modality for the facilitation of care of patients with progressive spasticity. The first patient suffers from Hypomyelination with Atrophy of the Basal Ganglia and Cerebellum (H-ABC). H-ABC is a rare condition, clinically characterized by progressive spasticity, dyskinesia and ataxia. The diagnosis is based on a typical Magnetic Resonance Imaging (MRI) pattern, consisting of hypomyelination and atrophy of the neostriatum and cerebellum[19]. The second patient suffers from progressive encephalopathy with brain atrophy most likely due to mitochondrial disease.

Case reports

Patient 1

The presented patient is the child of non-consanguineous Dutch parents without any known family history of neurological disease. The characteristics of this patient have been described in a previous publication (patient N° 6 in reference 19). He was born at term in 1983 and both pregnancy and birth were uneventful. Until the age of 2½ years his cognitive and motor development was normal. He then developed progressive spasticity and weakness, initially mainly involving the legs. His walking capability deteriorated, and he eventually became completely wheelchair-dependent at the age of 12. Until the age of 6 he had normal language and speech skills, but with increasing age he has had progressive dysarthria and a sensorineural hearing deficit. Finally, communication was only possible by means of gestures and pictograms.

Diagnostic tests at an early age were unrevealing. At 3 years of age a CT scan of the brain revealed no abnormalities, except for an enlargement of the lateral ventricles. EEG showed slowing of background activity; visually evoked potentials showed increased latencies. Metabolic screening of urine, blood and CSF showed no evidence of a metabolic disorder. A genetic evaluation for Pelizeus-Merzbacher disease was negative. The nature of the progressive leukencephalopathy remained unclear at that time.

The patient was referred to our department at the age of 18 because of increasing spasticity in the legs which interfered seriously with patient care. At that time, he was unable to speak and could only communicate with gestures (yes and no). Apart from deafness, the other cranial nerve functions were normal. The tendon reflexes in his arms and legs were symmetric and hyperactive. Examination of the arms showed dysmetria and dysdiadochokinesis and there was some spasticity of the triceps brachii muscle on both sides. Voluntary leg movements were not possible. There was severe spasticity of the legs and especially the hip-adductors. There were episodes of abrupt extensor movements in the trunk, hips and knees, which severely hampered his sitting position.

MRI showed profound hypomyelination of the cerebral hemispheres, as well as the pyramidal tracts at the level of the brain stem, absence of the putamen, and also atrophy of the caudate nucleus and the cerebellum (see Figure 1). On the basis of these findings he was diagnosed with H-ABC.

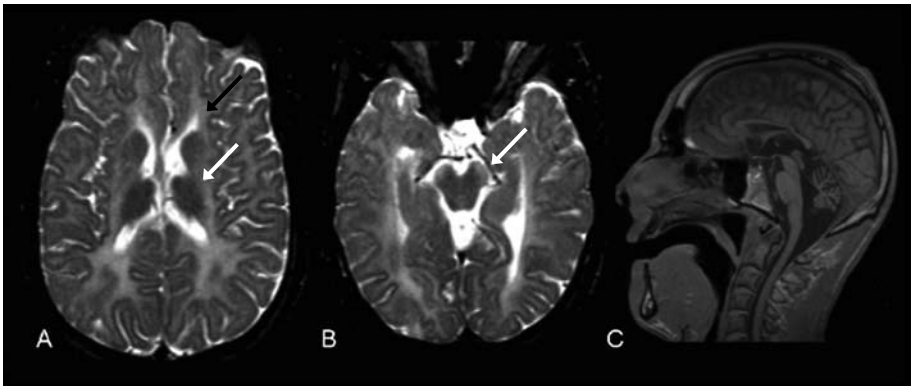
Because of his severe spasticity, which interfered with patient care, drug therapy with Dantrolene and Baclofen had been tried previously, without any success. We chose treatment with ITB. After trial treatment with intrathecal administration of Baclofen by lumbar puncture, an ITB pump was placed at the age of 19. Unfortunately, the patient developed a pouch infection, and the pump had to be removed 2 weeks after implantation. Retrospectively, the patient and his caregivers reported unsatisfactory improvement with ITB therapy.

The patient subsequently underwent laminotomy L2 to L5 and SDR of the spinal cord levels L2 to S1. The postoperative course was complicated by a urinary infection and recurrent migrating arthritis of unknown origin. However, the spasticity of the legs disappeared, resulting in a major improvement in patient care and sitting comfort in the wheelchair.

Because of recurrent back pain irradiating to the right leg, a CT scan of the cerebral spine was performed. This revealed compression of the spinal cord by a ventrally dislocated part of processus spinosus L1, which was removed surgically. The pain immediately decreased.

As expected, the clinical course remained progressive. The rigidity was not affected by the SDR, and was progressive. There were increasing posture-dependent extension spasms of the legs that were extremely uncomfortable and painful. These symptoms decreased after the administration of Baclofen orally. The cerebellar ataxia and pyramidal weakness were slowly progressive. Handling of the communication computer became impossible, and the patient lost the capability to manage his electric wheelchair independently. His orofacial motor skills decreased, resulting in problems with swallowing and episodes of aspiration. However, despite the symptoms of the progressive disease, the spasticity never recurred within a period of 3 years after SDR

FIGURE 1



MR images of patient 1 at the age of 22 years. A) Transverse T2-weighted MR image (3000/120) at the level of the basal ganglia. The white matter contains little myelin (black arrow) and the putamen is not visible (white arrow indicates the area where the putamen should have been). The thalamus and globus pallidus are of normal size. B) Transverse T2-weighted MR image (3000/120) at the level of the midbrain. The pyramidal tracts have high signal intensity (white arrow). C) Sagittal T1-weighted image (MP-rage 11.4/4.4). The cerebellum is severely atrophic and the corpus callosum is thin.

Patient 2

Patient 2 was born at a gestational age of 35 weeks by spontaneous delivery. His parents are of Turkish origin and are full cousins. His older sister has the same disease. His early motor development was delayed from early on. At the age of two years he could stand with support, but not sit or crawl. He could grab objects. Communication was possible by eye contact, gestures, and single syllable words. There was hypotonia and decreased stability of the trunk and head control. MRI, metabolic screening and chromosomal testing were normal at that time.

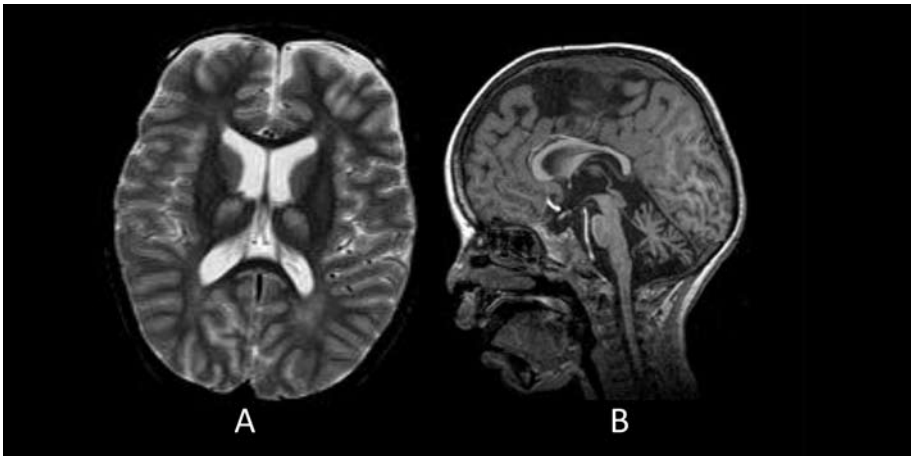
Subsequently, his motor skills and his cognitive function deteriorated with increasing hypertonia of the arms and legs. With four years he had developed serious spasticity in arms and legs. He had bilateral Babinski signs and hyperreflexia. Repeat MRI showed bilateral symmetrical signal abnormalities in the cerebral white matter and thalamus, in combination with cerebellar atrophy (see figure 2). A muscle biopsy was performed for mitochondrial studies. Deficiency of respiratory chain complexes II and IV was found. The basic genetic defect has not yet been identified.

The spasticity of the arms and legs increased over time. He developed extension and adduction spasms in the hips. Patient care became difficult and painful. He was repeatedly treated with Botulinum toxin A in the hip adductors, but the effects were short-lasting and unsatisfactory. He did not respond to oral Dantrolene and Baclofen, but Clobazam resulted in a slight reduction in tone.

At the age of seven, percutaneous radiofrequency rhizolysis was performed at the levels L1 to L4, with the aim to decrease the painful spasticity of the legs and to facilitate patient care. The effect was unsatisfactory. Although there was a moderate decrease of pain, spasticity and patient care did not improve. A serious spasticity was still present in the hip adductors, the hamstrings, the rectus femoris muscle and the triceps surae. SDR was performed at the age of eight years at the levels L2 to S1.

After SDR, leg spasticity disappeared completely. Extension spasms decreased dramatically. Sitting comfort in the wheelchair improved. Within a period of 3 years after SDR, leg spasticity defined as a velocity dependent stretch response did not recur. However, posturing with hip adduction recurred and patient care became painful again. These symptoms were completely reversible with oral Baclofen. Due to the progressive nature of the disease, spasticity in arms increased, leading to disabling contractures. Extension posturing of the trunk, which was interfering with sitting comfort, became a concern.

FIGURE 2



MR images of patient 2 at the age of 4 years. A) Transverse T2-weighted MR image (3000/120) at the level of the basal ganglia. The thalami contain signal abnormalities. B) Sagittal T1-weighted image (MP-rage 11.4/4.4). The cerebellum is severely atrophic.

Discussion

Outcome after SDR has mainly been studied in patients with CP, but SDR has also been performed in patients with spasticity due spinal cord injury[20,21] and multiple sclerosis[22,23]. Improvement of spasticity in the latter patients is less evident than in patients with CP. With regard to outcome after SDR in patients with a progressive neurodegenerative disease, very limited information is available[23].

The present case-reports support the concept that spasticity, defined as a velocity-dependent stretch response in a single joint, observed in patients with lesions of the corticospinal tract, improves or disappears permanently after partial interruption of the afferent fibres in the dorsal roots. Given the progressive nature of disease, it is not surprising that other symptoms than the leg spasticity, including arm spasticity, the cerebellar ataxia and general weakness, remained progressive after SDR.

In patient 1, however, spontaneous extensor spasms of the legs developed, despite SDR. Extensor spasms are mainly observed in patients with spinal cord injuries[24], but can also be seen in patients with multiple sclerosis[25] and neurodegenerative disease[26]. Salame et al. reported a long-term improvement in painful spasms after SDR in 16 out of 27 patients. In that cohort, however, some of the patients underwent other neurosurgical procedures, such as additional DREZotomy[23]. DREZotomy is a neurosurgical procedure which consists of the application of a microsurgical lesion of the dorsal root entry zones in the spinal cord (DREZ), and is mainly used to treat neuropathic pain[27]. Sindou et al. described a significant improvement in painful muscle spasms and abnormal flexion and/or extension postures after DREZotomy in patients with spastic paraplegia due to multiple sclerosis and trauma[28]. Clinical observations and experimental studies support the hypothesis that other pathophysiological mechanisms are responsible for spontaneous extensor spasms than for single-joint, velocity-dependent stretch response. Extension spasms seem to be pre-programmed stereotype responses to afferent inputs that include the hip and knee proprioceptors. It is assumed that in extensor spasms multi-segmental interneuronal pathways in the spinal cord are involved[29,30]. In SDR only the dorsal rootlets of the spinal nerves are transsected, leaving the spinal cord untouched. This may explain why the extensor spasms did not resolve after SDR in our patient, and why DREZotomy might have a better influence on extensor spasms than SDR.

In patient 2 extensor spasms were not a major concern. However, although spasticity of the hip adductors disappeared after SDR, the hip-adduction-posturing recurred. In patient 2 the major concern after SDR was the seriously progressive upper limb spasticity and the extension posturing of the trunk. These symptoms are consistent with the progressive nature of the disease.

Our conclusion is that SDR reduces spasticity in patients with a progressive neurological disease. The effect on the spasticity is permanent and irreversible. However, SDR does not necessarily have a beneficial effect on posture-dependent muscle spasms. The clinical goal of SDR in neurodegenerative disorders differs from that in spastic CP, where SDR is mainly performed to maintain walking ability. However, to improve patient care and sitting comfort, SDR proved to be effective. Other motor disturbances, such as ataxia, general weakness and spasticity of the arms are not influenced by SDR.

Before deciding to perform SDR, other treatment modalities, such as oral antispastic medication or intrathecal Baclofen, should be considered first. For patients suffering from a progressive neurodegenerative disease who do not respond to other treatments and for patients with a contra-indication for other treatments, SDR can be an option to reduce permanently spasticity.

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Chapter 4

Preoperative MRI findings and functional outcome after Selective Dorsal Rhizotomy in children with bilateral spasticity

Sebastian Grunt, Jules G. Becher, Petra van Schie, Willem J. R. van Ouwerkerk, Mazarin Ahmadi and R. Jeroen Vermeulen

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Abstract

Purpose

To identify MRI characteristics that may predict the functional effect of Selective Dorsal Rhizotomy (SDR) in children with bilateral spastic paresis.

Methods

We performed SDR in a group of 36 patients. The Gross Motor Functioning Measure-66 (GMFM-66) was applied before and after SDR. Available cerebral MRIs were retrospectively classified into three diagnostic groups: periventricular leucomalacia (PVL) [n=10], hydrocephalus, [n=2] and normal [n=6]. In patients with PVL we scored the severity of the MR abnormalities. We compared the changes in the GMFM-66 after SDR in the diagnostic groups. In patients with PVL we correlated the severity of the MR abnormalities with the changes in the GMFM-66.

Results

The mean follow up period was 5 years 4 months (range 1 year 1 month to 9 years). The best improvement in gross motor function was observed in patients with normal MRI, and the slightest improvement was observed in patients with hydrocephalus. The severity of the PVL did correlate with the GMFM-66-score before SDR but not with the functional effect of SDR.

Conclusion

We conclude that, with respect to gross motor skills, the improvements after SDR are good in patients with no MRI abnormalities. In the patients with hydrocephalus the improvements after SDR were insignificant. In patients with PVL the improvements were intermediate, and did not correlate with the degree of PVL.

Introduction

Spasticity is defined as a velocity-dependent increase in the tonic stretch response with excessive tendon jerk reflexes [1] and is caused by the reduction of inhibitory impulses on lower motor neurons. Spasticity can cause pain, muscle shortening and orthopaedic malformations, and it can severely interfere with functional abilities and gait pattern. Cerebral Palsy (CP) is the most frequent cause of spasticity in children. CP is a group of disorders of movement and posture due to a non-progressive lesion of the developing brain [2]. As a consequence of this definition, its causes are diverse.

Different treatment modalities are used to treat spasticity in children. Selective Dorsal Rhizotomy (SDR) is a neurosurgical treatment that is mainly performed at lumbar level in patients with bilateral spastic paresis. By incomplete transsection of the (sensory) posterior lumbosacral rootlets, SDR reduces the excitatory input from the lower limbs that enters the spinal cord. In children with spastic CP, SDR leads to a more normal gait pattern [3–6], better performance of the activities of daily life [7, 8] and improvement in gross motor function [6–13]. Data on functional outcome after SDR in patients with other conditions than spastic CP are limited, though promising [14]. Detailed information about the clinical characteristics of the selected children, other than spasticity alone, is often lacking.

Medical history and clinical examination does not always provide sufficient evidence to establish the diagnosis of the underlying disorder in patients with CP. The Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurology Society both advise Magnetic Resonance Imaging (MRI) of the brain for all CP patients [15]. In patients with bilateral spastic CP, the most common MRI abnormality is periventricular leucomalacia (PVL), which is characterised by a damage of the periventricular white matter [16–19]. Grey matter lesions, early developmental abnormalities of the central nervous system and abnormalities of the cerebrospinal fluid space, such as hydrocephalus are less commonly found [17–19] and in some children with bilateral spastic CP no abnormalities are found with MRI [16–19]. In patients with normal MRI findings and bilateral spastic paresis, genetic causes or spinal cord involvement should be considered.

The goal of this study was to identify possible relationships between the MRI findings with the level of gross motor function of the patient before SDR and the change in functioning after SDR. For this purpose we retrospectively analyzed the MRIs of the patients who underwent SDR in our clinic.

Methods and Materials

Subjects

This study was part of the project: "Investigation of long term effects of SDR in children with spastic diplegia" and was approved by the medical ethical committee of the VU University Medical Center, Amsterdam, the Netherlands (project Nº 2006/105). Informed consent was given by the parents of all participants. The study population consisted of all the patients who underwent SDR in our clinic between January 1998 and December 2007. The first nine patients have been described in a previous report [8]. The patients were selected for SDR according to the criteria shown in Table 1. In our previous study we only included patients with bilateral spastic CP and documented PVL [8]. In the present study we included patients with non-progressive spasticity, due to different diagnoses. In our analysis we included data on patients who underwent pre- and post-operative Gross Motor Function Measure (GMFM-66) analysis with a follow up period of at least 12 months and of whom MRI was available for review.

Procedure

SDR was performed by the same neurosurgeon (WVO) in all children. Dorsal roots L2-S1 were exposed and separated into different rootlets after laminotomy L2-L5 and opening of the dura. Transsection of the rootlets was performed after electrostimulation, according to palpable muscle contraction and EMG response. At most, 50% of the rootlets were trans-sected on one level. To prevent sexual and bladder disturbances, rootlets/fascicles showing electrical response after stimulation of the penis/the clitoris were spared. Post-operative rehabilitation included intensive physical therapy for 12 months.

TABLE 1 **Selection criteria for Selective Dorsal Rhizotomy in the VU University Medical Centre**

– Bilateral spasticity of the lower extremities interfering with walking performance
– Presence of spasticity (defined as velocity-dependent resistance to passive stretch) in at least six muscle groups of the lower limbs
– Sufficient force in the quadriceps femoris muscle (squatting at least 7 times) and the hip extensors (kneel with extended hips, support for balance allowed)
– Absence of structural orthopaedic deformities or contractures at hip, knee or ankle
– Presence of moderate to good selective motor control in the lower limbs
– Gross Motor Function Classification System (GMFCS) level I, II or III
– Good support from parents and rehabilitation setting

Neuroimaging

The MRIs were assessed at two different points in time by two investigators – one neuropaediatrician (8 years of experience in the assessment of MR neuroimaging [RJV]) and one paediatrician with specialization in paediatric neurorehabilitation (2 years of experience in the assessment of MR neuroimaging [SG]). Imaging was classified into three diagnostic groups according to the following criteria: periventricular leucomalacia (PVL) (increased signal intensity of the periventricular white matter in T2 weighted imaging and/or FLAIR; other abnormalities optional); hydrocephalus (signs of ventricular dilatation and increased intracranial pressure); normal MRI (see figure 1a-i). In one patient the MR abnormalities could not be classified into one of the three diagnostic groups and he was excluded from further data-analysis (patient N 19, table 2; his MRI showed delayed myelinisation and slightly enlarged ventricular size, but no other white matter abnormalities and no signs of increased intracranial pressure). In the patients with PVL we graded the severity of their MRI abnormalities, based on the work of Cioni et al. [20]. The following items were assessed: ventricular size, evidence and extension of white matter signal intensity, evidence and extension of white matter loss, thinning of the corpus callosum, dimension (size) of sub-arachnoidal space, evidence and size of cysts, and presence of grey matter abnormalities. We also investigated whether white matter loss was present occipitally and/or frontally. The items were scored on a 3-point scale, with a score of 3 indicating the most severe MRI abnormalities. The scores were summed to obtain a total score for each child (minimal score = 7, maximal score = 21). Intra- and inter-observer agreement was assessed for all items. If the two investigators did not agree, the classification and/or scoring were determined by a consensus.

Outcome measure

All patients had a detailed pre- and post-operative clinical evaluation, including spasticity assessment, range of motion of single joints, gait-analysis and the GMFM. In this study we only report the results of the GMFM. The GMFM is a criterion-referenced observational measure that was developed to assess children with CP. The validity, reliability and responsiveness of the GMFM have been demonstrated in a population of patients similar to the participants in the present study [21]. For our data-analysis we used the GMFM-66 version, which assesses 66 items covering five gross motor dimensions (lying and rolling/crawling and kneeling/standing and walking/running/jumping) and is elaborated to a numerical scale ranging from 0 to 100.

As outcome parameters we used the mean of all post-operative GMFM-66 scores from 1 year after surgery. The changes after SDR were expressed as the difference between GMFM-66 score and the pre-operative GMFM-66 score (Delta-GMFM-66).

Statistical analysis

For the reliability analysis of the MRI scoring we used Kappa statistics. Agreement strengths for the Kappa values were classified according to Landis and Koch [22]. For the comparison of the outcome in patients with different MRI characteristics we used non parametric tests (Kruskal-Walis test for group comparison, post hoc analysis was performed by the Mann-Whitney test when the level of significance was reached). Correlations were assessed with the Spearman's rank correlation coefficient. For the statistical analysis we used SPSS® version 14.0 for Windows. Level of significance was set at 0.05.

Results

Clinical characteristics and level of functioning

Thirty-six patients underwent SDR of whom 32 patients had a brain MRI performed before SDR, and 26 MRIs were available for review. Of the patients with available MRI, four did not have any pre-operative GMFM-66 assessments. Three patients had a GMFM-66 assessment before SDR, but a follow up period of less than 12 months. The detailed patient characteristics of the remaining 19 patients are summarized in Table 2. The mean number of GMFM assessments per patient was 4.9 (standard deviation [SD] 3.5, range 1 to 12), and the mean follow up period was 5 years 4 months (SD 2 years 9 months, range 1 year 1 month to 9 years). The mean age at the time of the operation was 6 years 10 months (SD 1 years 6 months, range 5 years 9 months to 10 years 1 month). The age at the time of SDR did neither correlate with the preoperative GMFM-66 (rho = 0.328, p = 0.184) nor with the postoperative GMFM-66 scores (rho = 0.336, p = 0.173) and the improvement after SDR (rho = –0.049, p = 0.847). We found no difference in the Delta-GMFM-66 score between boys and girls.

TABLE 2 **Summary of the characteristics of the patients included in the study**

Case	Gender	GA (wks)	BW (g)	Age SDR years (mts)	Follow-up years (mts)	MRI Classification	Diagnosis	GMFM-66 before SDR	GMFM-66 (mean) after SDR
1	F	30	2000	5 (7)	8 (5)	PVL	PVL	50.85	53.29
2	F	40	2500	5 (7)	9 (0)	PVL	PVL	47.68	49.66
3	F	40	3450	5 (8)	8 (11)	PVL	PVL	54.38	55.71
4	M	30	1650	6 (8)	8 (11)	PVL	PVL	46.91	48.32
5	F	28	1285	4 (11)	7 (10)	PVL	PVL	50.32	54.75
6	M	26	1000	5 (3)	7 (10)	PVL	PVL	64.98	67.82
7	M	32	2510	5 (1)	6 (7)	PVL	PVL	54.15	60.38
8	M	33	1265	8 (9)	5 (1)	PVL	PVL	47.09	55.92
9	M	26	870	6 (11)	3 (11)	PVL	PVL	50.09	54.32
10	M	26	780	5 (4)	3 (0)	PVL	PVL	52.32	62.98
11	F	41	3060	8 (4)	5 (2)	Hydrocephalus	Congenital Hydrocephalus	55.62	56.35
12	M	27	1020	10 (1)	4 (5)	Hydrocephalus	Congenital Hydrocephalus	68.86	68.69
13	M	40	3300	5 (9)	7 (2)	Normal	Unknown	65.33	67.31
14	M	34	3155	6 (11)	3 (0)	Normal	Spinal Process	76.75	85.15
15	M	38	4280	8 (0)	2 (7)	Normal	Unknown	82.99	89.70
16	F	40	3875	8 (10)	1 (7)	Normal	HIV-Encephalo(myelo)pathy	73.63	80.46
17	M	39	4040	6 (7)	2 (0)	Normal	Unknown	65.98	78.38
18	M	40	3460	6 (5)	1 (1)	Normal	Unknown	73.63	81.93
19	M	37	3400	3 (11)	7 (1)	Not classified	Laurence-Moon Syndrome	47.26	83.01

Abbreviations used: F = female, M = male; GA = gestational age, BW = birth weight, SDR = selective dorsal rhizotomy, GMFCS = Gross Motor Function Classification System, GMFM = Gross Motor Function Measure, PVL = periventricular leucomalacia

Neuroimaging

We diagnosed PVL in 10 patients, hydrocephalus in 2 patients and no MRI abnormalities in 6 patients. The classification of MRIs did not differ between the two observers. With respect to the grading of the MR abnormalities, the intrarater agreement was perfect for the frontal and the occipital white matter loss, the cysts, the thinning of the corpus callosum, the subarachnoidal space and the grey matter abnormalities (Kappa = 1.0) and almost perfect for the ventricular size (Kappa = 0.90) and the white matter signal intensities (Kappa = 0.87). The interrater agreement was considerably less than the intrarater agreement. It was perfect for the subarachnoidal space and the grey matter abnormalities (Kappa = 1.0), almost perfect for the ventricular size (Kappa = 0.90), substantial for the occipital white matter loss (Kappa = 0.74) and moderate for the frontal white matter loss (Kappa 0.45). Kappa statistics could not be calculated for the white matter signal intensities, the cysts and the thinning of the corpus callosum as only one of the two observers graded severe abnormalities (according grade 3 on the point scale). With respect to the white matter signal intensities and the thinning of the corpus callosum the observers disagreed in 25% of the scorings, with respect to the cysts there was a disagreement in 36%.

The outcome parameters in patients with different MRI classification are summarized in Table 3 and in figure 2. The pre-operative GMFM-66 scores were significantly higher in patients with normal MRI than in patients with PVL ($p = 0.001$). Patients with hydrocephalus had intermediate scores and did not differ significantly from the other groups. In the follow-up measurements the mean GMFM-66 was the highest in patients with a normal MRI (group difference $p = 0.003$, difference between PVL and normal $p = 0.002$). The patients with normal MRI also made the best post-operative improvements. Almost no improvement was observed in the patients with hydrocephalus (group difference $p = 0.030$, difference between PVL and hydrocephalus $p = 0.032$, difference between hydrocephalus and normal MRI $p = 0.046$). There was a significant group difference in age at the time of the operation ($p = 0.028$). Patients with normal MRI and patients with hydrocephalus were older at the time of the operation than patients with PVL – for the patients with normal MRI this difference was significant ($p = 0.044$).

The scoring of the MRI abnormalities in the patients with PVL are summarized in table 4. Two patients with PVL did not show ventricular enlargement, and eight patients had moderate ventricular enlargement. All patients had moderate white matter signal intensity. Nine patients had moderate occipital white matter loss and one patient had moderate frontal white matter loss. Small cysts were found in one patient, and large cysts were found in two. Four patients had no thinning of the corpus callosum, and six patients had moderate thinning of the corpus callosum. None of the patients had grey matter abnormalities or an enlargement of the sub-arachnoidal space.

The mean total score was 10.8 (SD 1.1, range 8 to 12). The total score did not correlate with any of the outcome parameters. The ventricular size showed a significant correlation with the pre-operative GMFM-66 score (rho = –0.696, p = 0.025). However, none of the items in the scoring correlated with the post-operative changes of the GMFM-66.

FIGURES 1a – 1i **MR imaging of three patients with bilateral spastic paresis undergoing SDR. Fig. 1a, 1d, 1g: Midsagittal T1 weighted images. Fig. 1b, 1e, 1h: Transversal T2 weighted images at the level of the centrum semiovale. Fig 1g, 1h, 1i: Transversal T2 weighted imaging at the level of the basal ganglia. Fig. 1a–1c: MR images classified as "normal". Fig. 1d–1f: MR images classified as periventricular leucomalacia, showing thinning of the corpus callosum involving the total body (Fig. 1a), bilaterally increased periventricular white matter signal intensity (Fig. 1e), a slight ventricular enlargement and a loss of the occipital white matter (Fig. 1f). Fig. 1g-1i: MR images classified as hydrocephalus. The lateral ventricles and (Fig. 1i), the third ventricle and the fourth ventricle are enlarged.**

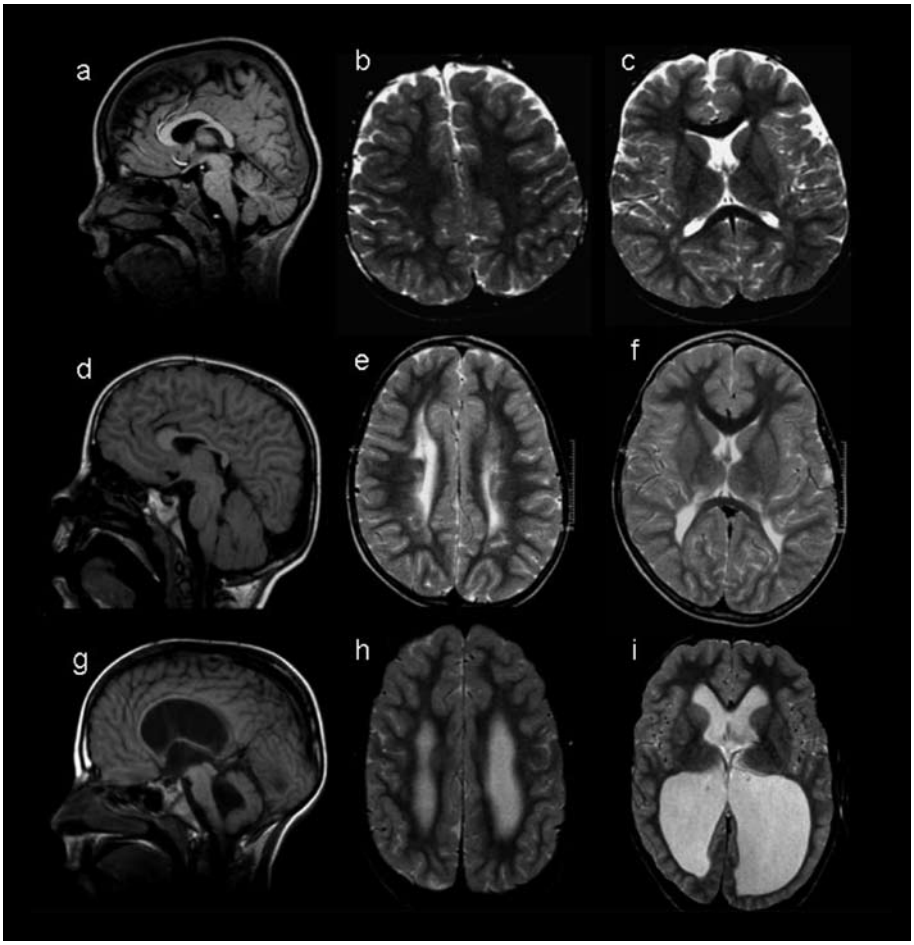

TABLE 3 **Differences of gross motor outcome in with patients with different MRI classification**

	Normal MRI (n=6)			PVL (n=10)			Hydrocephalus (n=2)			Total (n=18)			P value
	Mean	SD	Range	Mean	SD	Range	Mean	SD	Range	Mean	SD	Range	
GMFM-66 before SDR	73.1†	6.7	65.3–83.0	51.9†	5.3	46.9–65.0	62.2	9.4	55.6–68.9	60.1	11.5	46.9–83.0	0.002
Mean GMFM-66 after SDR	80.5†	7.6	67.3–89.7	56.3†	5.9	48.3–67.8	62.5	8.7	56.3–68.7	65.1	13.0	48.3–89.7	0.003
Delta GMFM-66	7.4‡	3.4	2.0–12.4	4.4§	3.2	1.3–10.7	0.3‡/§	0.6	0.2–0.7	5.0	3.7	-0.2–12.4	0.030

Abbreviations used: MRI = Magnetic Resonance Imaging, PVL = periventricular Leucomalacia, SD = Standard Deviation, GMFM = Gross Motor Function Measure
† Significant difference with p<0.05 (Mann Whitney test) between Normal MRI and PVL,
‡ Significant difference with p<0.05 (Mann Whitney test) between Normal MRI and Hydrocephalus
§ Significant difference with p<0.05 (Mann Whitney test) between Hydrocephalus and PVL

FIGURE 2 **Box Plots of the pre-operative GMFCM-66 (white boxes) and the mean values of all postoperative GMFM-66 measurements (grey boxes) in patients with different MRI classification. Note that the largest improvements after SDR were observed in patients with normal MRI and that the patients with hydrocephalus did not improve after SDR.**

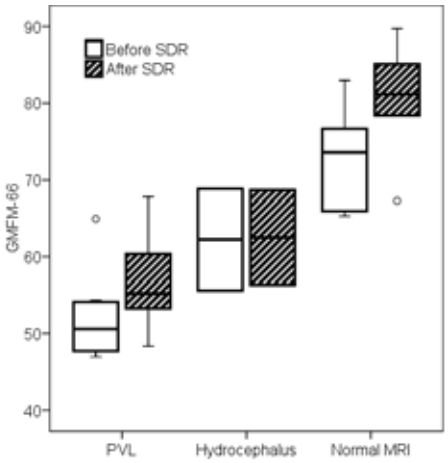

TABLE 4 **Scoring of the MR-abnormalities in patients with periventricular leucomalacia (n=10)**

	Normal	Moderate	Severe
Ventricular Size	2 (20%)	8 (80%)	0 (0%)
White Matter Signal Intensity	0 (0%)	10 (100%)	0 (0%)
White Matter Loss	1 (10%)	9 (90%)	0 (0%)
Thinning of the Corpus Callosum	4 (40%)	6 (60%)	0 (0%)
Cysts	7 (70%)	1 (10%)	2 (20%)
Grey Matter Abnormality	10 (100%)	0 (0%)	0 (0%)
Enlargement of the Subarachnoidal Space	10 (100%)	0 (0%)	0 (0%)
	Mean	**SD**	**Range**
Total Score	10.8	1.1	8 – 12

Discussion

The aim of this study was to identify a possible relationship between MR characteristics and the level of functioning before and after SDR in patients with bilateral spastic paresis. We classified the MRIs in three MRI categories: PVL, hydrocephalus and normal MRI, and compared the post-operative improvement in the GMFM in the three groups. The best results were found in the group with normal MRI, and the poorest results were found in patients with hydrocephalus. The poor outcome of patients with hydrocephalus could be a consequence of persistently elevated intracranial pressure, which can be observed in the absence of characteristic symptoms in children with CP [23]. In the present study both patients with hydrocephalus (patient 11 and patient 12) underwent surgical correction. Patient 11 had a ventriculo-peritoneal shunt placed early during childhood. Patient 12 was diagnosed with hydrocephalus a few months before SDR and he underwent third-ventriculostomy. Unfortunately, there was no follow-up imaging available for either of these patients, and although neither of them showed any clinical evidence of elevated intracranial pressure, it cannot be ruled out definitively. Previous studies found greater post-operative improvements after SDR in children with less motor impairment [7, 13]. In contrast, in the three randomised controlled trials comparing functional outcome after SDR, the poorest results were found in the study which included children with the best pre-operative gross motor skills [9, 10]. In the present study, the patients with normal MRI had much better pre-operative gross motor skills than the patients with PVL and hydrocephalus. We found no correlation between the pre-operative GMFM score and its improvement post-operatively. McLaughlin et al. found an inverse correlation between the age at the time of the operation and the post-operative changes in the GMFM [9]. This could be explained as a consequence of faster spontaneous motor de-

velopment in early during childhood. However, in our study not only the children with hydrocephalus (who showed the poorest outcome) but also the children with normal MRI (who showed the best outcome) were older than the patients with PVL when SDR was performed. There was no correlation between the age at the time of SDR and outcome.

In the patients with PVL we graded the severity of the MR abnormalities and correlated them with the gross motor abilities. For this purpose we used a grading scale that has previously been used to describe MR abnormalities in patients with spastic CP [20, 24]. We performed a reliability analysis of the various items included in this grading scale. The agreement between investigators was substantially worse than in the Cioni et al. study, in which two experienced raters scored the MRIs [20]. In the present study the inter-observer reliability was notably poor for the assessment of the cysts. Several explanations should be considered: it was difficult to detect cysts in an area with extensive gliosis, there is a difference between the level of experience with brain MRI in the two investigators (2 years vs. 8 years). Only the amount of ventricular enlargement correlated with the pre-operative gross motor abilities. Correlations between ventricular size and gross motor abilities have been described previously in patients with spastic CP [25]. In contrast to the findings of previous studies [20, 24, 26] we did not find a correlation between gross motor skills and the total MRI score/the thinning of the corpus callosum. However, the present study was limited to ambulatory patients with GMFCS levels I to III and patients with more severe motor handicaps where more severe brain abnormalities could be expected on MRI have not been studied. No correlations were found between the severity of the brain anomalies and the outcome after SDR.

The outcome assessment in the present study consisted in the GMFM-66 which – according to the International Classification of Functioning, Disability and Healths (ICF) [27] – assesses the domain of activity. However, the other two domains of the ICF – body structure and participation – have not been evaluated. One other limitation of the present study was the small sample size. Unfortunately, we did not have access to all MRI scans and not all patients had a pre-operative GMFM assessment. We could only include data on a small number of patients in our analysis. Notably the group with hydrocephalus was small and comprised only 2 patients. Therefore, the association between the type of brain lesion and the outcome after SDR needs to be proved in future studies with larger study samples, including the other ICF domains – body structure and participation.

Conclusion

We found significant differences in the post-operative changes in the GMFM-66 in patients with different brain MRI abnormalities. The largest postoperative improvement was observed in patients with normal MRI, and the poorest outcome was observed in patients with hydrocephalus. In patients with PVL we could not detect any relation between MRI abnormalities and the postoperative improvement in of gross motor function after SDR. We conclude that MRI of the brain can provide additional information for the selection of patients for SDR. However, the degree of PVL does not provide information about the degree of improvement in gross motor function after SDR.

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Chapter 5

Reproducibility and validity of video screen measurements of gait in children with spastic cerebral palsy

Sebastian Grunt, Petra J. van Kampen, Marjolein M. van der Krogt, Merel-Anne Brehm, Caroline A.M. Doorenbosch and Jules G. Becher

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Abstract

Purpose
To determine the reproducibility and validity of video screen measurement (VSM) of sagittal plane joint angles during gait.

Methods
17 children with spastic cerebral palsy walked on a 10m walkway. Videos were recorded and 3d-instrumented gait analysis was performed. Two investigators measured six sagittal joint/segment angles (shank, ankle, knee, hip, pelvis, trunk) using a custom-made software package. The intra- and interrater reproducibility were expressed by the intraclass correlation coefficient (ICC), standard error of measurements (SEM) and smallest detectable difference (SDD). The agreement between VSM and 3d joint angles was illustrated by Bland-Altman plots and limits of agreement (LoA).

Results
Regarding the intrarater reproducibility of VSM, the ICC ranged from 0.99 (shank) to 0.58 (trunk), the SEM from 0.81° (shank) to 5.97° (trunk) and the SDD from 1.80 (shank) to 16.55° (trunk). Regarding the interrater reproducibility, the ICC ranged from 0.99 (shank) to 0.48 (trunk), the SEM from 0.70° (shank) to 6.78° (trunk) and the SDD from 1.95° (shank) to 18.8° (trunk). The LoA between VSM and 3d data ranged from 0.4 +/- 13.4° (knee extension stance) to 12.0 +/- 14.6° (ankle dorsiflexion swing).

Conclusion
When performed by the same observer, VSM mostly allows the detection of relevant changes after an intervention. However, VSM angles differ from 3d-IGA and do not reflect the real sagittal joint position, probably due to the additional movements in the other planes. When VSM is used for clinical decision-making, measurement errors must be considered, particularly when VSM is performed by different observers. For clinical questions as well as for scientific purposes three dimensional recordings are preferrable.

Introduction

Three-dimensional instrumented gait analysis (3d-IGA) is considered to be the gold standard for the assessment of gait abnormalities in patients with movement disorders [1]. It provides an accurate description of gait kinetics, gait kinematics and muscle activation, and can help to plan and evaluate treatment in patients with gait disorders [2,3]. The use of 3d-IGA, however, has some disadvantages: It requires special equipment, is time consuming, is dependent on patient compliance, and it is not always available. Observational gait analysis (OGA) has been proposed as an alternative to 3d-IGA [4]. In OGA an observer visually assesses the gait pattern, mostly with the aid of video recordings. Various OGA scales have been developed and modified [4–6]. For the detection and quantification of gait abnormalities these OGA scales rely on subjective ordinal scales that describe the gait abnormalities in different joints and planes. The reproducibility of OGA differs considerably between studies and between the single items within one OGA scale [4–8]. Kappa values ranged from –0.04 to 0.91 for intrarater repeatability and from 0.13 to 0.94 for interrater repeatability [4–7]. With respect to the agreement between OGA and 3d-IGA, the kappa values ranged from –0.11 to 0.94 [6,7,9] and the agreement ranged from 43% to 83% [4].

OGA scales provide a good general impression on the severity of a patient's gait deviation [10], but do not provide specific information on the gait kinematics of a single joint. In the evaluation of gait abnormalities one often relies on more detailed kinematic measurements. Different software programs allowing a 2-dimensional joint angle measurement on video screen (i.e. video screen measurement: VSM) have been developed [11–14]. Two studies evaluated the validity of VSM during gait in disabled patients [6,14]. In both studies the measurements were restricted to the knee joint. Kiernan et al. showed an overall level of agreement of 2 +/–13° between the 3d-IGA and VSM technique [14]. In the study of Dickens et al. the intraclass correlation coefficients (ICC) for the VSM versus the 3d-IGA measurements ranged from 0.44 to 0.90, and the ICC for the knee angle measurements of two different observers ranged from 0.95 to 0.96 [6]. The intrarater reproducibility was notinvestigated. The aim of the present study was to assess the reproducibility of measuring sagittal kinematics during gait in six different joints and segments using the VSM technique [13]. In order to assess whether this tool can be used for clinical decision making and/or as an outcome measure of clinical interventions the following questions were investigated: A: How reproducible are the measurements for repeated assessments by the same observer? B: How reproducible are the measurements for assessments by two different observers? And C: How much do the measurements differ from 3d-IGA?

Methods

Participants

17 children with spastic CP participated in the study. Their mean age was 8.9 years (SD 2.1 years, range 6–12 years). Thirteen children were diagnosed with bilateral and four with unilateral spastic CP. Of the four children with unilateral CP, three were affected on the right side and one on the left side. All children were able to walk without assistive devices (Gross Motor Function Classification System level I or II) [15]. Participants did not have any prior orthopedic surgery, rhizotomy or baclofen treatment or any treatment with botulinum toxin within the previous 16 weeks.

Procedure

Data used in the present study were obtained from the study by Van der Krogt et al. [16]. This study was approved by the Medical Ethics Committee of the VU University Medical Center. The subjects walked barefoot along a 10m walkway at a self-selected comfortable speed. Video recordings of the right leg were taken in the sagittal and frontal plane. Three- dimensional (3d) kinematic data were simultaneously recorded from the trunk, pelvis, thigh, shank and foot, using a motion capture system (Optotrak, Northern Digital, Waterloo, Canada). A technical cluster of three markers was attached to each segment. Anatomical landmarks were indicated in order to anatomically calibrate the technical cluster frames [17]. Two trials per subject, collected during two separate sessions, were used. One trial was randomly selected for each session, and for each of these two trials one representative stride was chosen for the assessment. Since the aim of this study was to assess the repeatability of the measurement, and not the variability within one individual subject, each of these two trials was considered as a separate case.

Data analysis

For the VSM, a custom-made, open-source software package, which allows to synchronously observe gait parameters and video recordings was used (the MoXie Viewer®, www.smalll.nl) [13]. The speed of the videos can be freely selected, including slow motion and freeze frame techniques. The software includes a graphical multi-goniometer that allows measuring and displaying six sagittal angles (see Figure 1). The sagittal joint/segment angles were determined as follows. Ankle: angle between shank and hindfoot; shank: angle between shank and ground; knee: angle between shank and thigh; hip: angle between thigh and pelvis; pelvis: angle between pelvis and ground; trunk: angle between trunk and pelvis.

Two investigators (one paediatrician [observer 1] and one physiatrist [observer 2]) measured all sagittal angles in each phase of the gait cycle (initial contact, loading response, mid stance, terminal stance, pre-swing, initial swing, mid swing and terminal swing) within the representative strides. Both investigators had the same level of experience (working in the field of pediatric neurorehabilitation for five years, experience in gait analysis for two years). For each measurement, one specific video frame was defined per phase. To assess the intrarater repeatability observer 1 reassessed the videos three weeks later. In order to compare the VSM and 3d-IGA technique observer 1 measured the following sagittal angles within the same selected strides: peak ankle dorsiflexion in stance, peak knee extension in stance, peak hip extension in stance, peak ankle dorsiflexion in swing, peak knee flexion in swing and peak hip flexion in swing.

Three-dimensional kinematic data were analysed using custom-made, open-source software (www.BodyMech.nl, Matlab®, The Mathworks). Joint and segment angles and angular velocities were calculated following the CAMARC anatomical frame definitions [18]. The 3d data were synchronised with the video data, and imported into the MoXie viewer® software in order to view the video frames and 3d data simultaneously. Three dimensional gait data were obtained synchronously with video measurements, but the two investigators did not have access to the 3d data until after the video screen measurements had been performed.

Statistical analysis

Reproducibility was assessed for each joint/segment and for each phase of the gait cycle according to the Generalizability theory, which is based on analysis of variances (ANOVA) [19]. The components of variance estimated with this analysis included the intersubject variance (case); the variance related to repeated sessions (occasion); the variance related to observers (investigator) and the error variance (error). Based on these variance components, the Intraclass Correlation Coefficient (ICC), the standard error of measurement (SEM) and the smallest detectable difference (SDD) were calculated according to the equations presented in Table 1 [19,20]:

$$ICC_{intrarater} = case/(case+occasion+error)$$

$$ICC_{interrater} = case/(case+occasion+investigator+error)$$

$$SEM_{intrarater} = \sqrt{(occasion+error)}$$

$$SEM_{interrater} = \sqrt{(occasion+investigator+error+[occasion*investigator])}$$

$$SDD_{intrarater} = 1.96*SEMintrarater*\sqrt{2}$$

$$SDD_{interrater} = 1.96*SEMinterrater*\sqrt{2}$$

The agreement between VSM and 3d-IGA was calculated by the 95% limits of agreement and illustrated by Bland-Altman Plots. The Bland-Altman plot is a graphical method that can be used to compare two measurements techniques. Within the plot, the differences between the two techniques are plotted against the averages of the two techniques. Horizontal lines are drawn at the mean difference, and at the 95% limits of agreement (LoA), which are defined as the mean difference plus and minus 2 times the standard deviation of the differences [21]. For the statistical analysis SPSS Version 14.0 for Windows was used.

Results

Thirty trials were available for the assessment. Fourteen subjects performed walking trials at two different sessions, 3 subjects performed walking trials at one session. One video assessment could not be evaluated for technical reasons. Table 1 shows the reproducibility results for measurements of the same observer. Table 2 shows the reproducibility results for measurements of two different observers. The reproducibility varied per phase of the gait cycle and per joint/segment. For repeated measurements of the same observer it was best for the shank (ICC=0.97, range 0.92–0.99) and poorest for the trunk tilt (ICC=0.74, range 0.58–0.85). The mean SEM ranged from 1.6° (shank; range 0.6°–5.9°) to 3.1° (trunk tilt; range 2.2°–6.0°) and the mean SDD ranged from 4.5° (shank; range 1.8°–16.5°) to 8.5° (trunk tilt; range 6.1°–16.5°). For measurements of two different observers the reproducibility was also best for the shank (ICC=0.96, range 0.93–0.99) and poorest for the trunk tilt (ICC=0.58, range 0.48–0.72). The mean SEM ranged from 1.2° (shank; range 0.7–1.5°) to 4.4° (trunk tilt; range 3.1°–6.8°) and the mean SDD ranged from 3.3° (shank; 1.9°–4.3°) to 12.1° (trunk tilt; 8.6°–18.8°). To determine the validity, the agreement between the VSM and 3d-IGA technique was determined with the Bland-Altman plots (Figure 2). There was a wide range of variation between the measurements. Agreement was poorest for the peak ankle dorsiflexion in swing (LoA=12 +/–14.6 degrees) and best for the peak knee flexion in swing (LoA=–1.4 +/–11 degrees). The mean difference between the VSM and 3d-IGA technique was 5.7° for peak ankle dorsiflexion in stance (Fig 2a) and 12° for peak ankle dorsiflexion in swing (Fig 2b) which means that the 3d measurements of the ankle angles systematically showed more plantar flexion than the VSM.

Discussion

In clinical practice video based gait analysis is an important alternative to 3d instrumented gait analysis. However, only little information is available on the reproducibility and validity of this technique. The present study therefore examined the reproducibility and validity of video screen measurement (VSM) of six different joints/segments during each of the pre-defined 8 phases of the gait cycle in children with spastic cerebral palsy.

The reproducibility of the measurements has been described by different reproducibility measures: The SEM, the SDD and the ICC. With respect to the SEM, McGinley stated that a measurement error of 2° or less is acceptable, and a measurement error ranging from 2 to 5° can be regarded as reasonable, but may require consideration in the data interpretation [22]. In the present study measurement errors of less than 2° were found for the shank. For all the other joints/segments the measurement errors ranged between 2° and 7° and varied between the phases of the gait cycle and the joints/segments. These measurement errors are comparable to the measurement errors that are found in 3d IGA [22]. These errors must be taken into account when VSM is performed.

In the clinical settings, kinematic gait data are of particular interest when evaluating changes after an intervention. According to the results of the present study VSM allows detecting a change between 2 and 19° – depending on the joint/segment angle and the phase of the gait cycle. For a meaningful interpretation of this change, an assumption is required of what level of change can be considered as clinically relevant [23]. However, it is beyond of the scope of this article to define the limits of clinical relevance. Changes after an intervention must always be interpreted in the context of multiple factors, such as the type of intervention, the joint/segment concerned and outcome measures other than the kinematic gait data. In our opinion a change of 10° or morecan be considered as relevant in the majority of the cases. With respect to VSM, this implies, that the same observer could detect such a change in all joints/segments (with the exception of the shank in loading response and the trunk at initial contact). When two different observers perform the measurements, the SDD is more than 10° in the ankle (terminal stance, midswing), hip (midstance, preswing, midswing), pelvis (midswing) and the trunk (initial contact, loading response, midstance, midswing and terminal swing). For the assessment of treatment outcomes the same observer should perform VSM before and after the intervention.

In contrast to the SEM and SDD, the ICC does not express the reproducibility in the same unit as the original data. The ICC takes into consideration the variability of the original data [23]. This explains why the ICC can be relatively low in the presence of small measurement errors. In the literature, the ICC is commonly used as a measure of reproducibility, and therefore it was calculated in the present study to allow a comparison with previous findings. For the knee angle the results of the present study were comparable to the results that have been found by Dickens et al. [6]. In the present study the ICC for the interrater reproducibility ranged from 0.88 to 0.97 whereas in the study of Dickens et al. The ICC ranged from of 0.95 to 0.96 [6]. In line with Dickens et al. [6], our findings support that joint and segment angular data from VSM may provide a more repeatable assessment than the categorical data that are commonly used in observational gait scales.

The usefulness of a measurement tool is not only dependent on its repeatability, but also on its validity. In the present study the validity of VSM was evaluated by comparing its measurement outcomes to the corresponding outcomes derived with 3d-IGA. Since small synchronization errors between video data and 3d data can lead to large differences, only peak joint angles in three different joints (ankle, knee and hip) were assessed and compared between the two techniques. When measuring peak joint angles instead of joint angles in predefined video frames, bias due to synchronization error is avoided. Similarly to previous studies [6,15] the LoA showed a large variation. For the ankle angles, a systematic difference between the two techniques was found. VSM underestimated the amount of plantarflexion compared to 3d kinematic data (see Figure 2). This disagreement can be explained by the differences of anatomical definitions within the two techniques. In the VSM technique the ankle angle was defined as the angle between the tibia and the line of the foot sole at the level of the hind foot. In the 3D-model [18] the foot reference frame (defined by the upper ridge of calcaneus and MTP 1–5) was in a few degrees plantar flexion in the anatomical position. These systematic differences in definition can be taken into account when analyzing the data, and therefore do not necessarily represent a validity problem. However, differences in agreement for the ankle angles may also have been influenced by hindfoot, midfoot and forefoot position in the coronal and transverse planes. Abnormalities of the foot segment in these planes can make sagittal measurements on video difficult, and this may also impact the accuracy 2d sagittal measures. No systematic measurement error was found for the hip and knee angles (see Figure 2), and the differences between VSM and 3d-IGA are hypothesized to be mainly due to movements of the hip and the pelvis in the transverseal/frontal plane. VSM is restricted to the 2D sagittal plane and movements in other planes cannot be measured. Therefore VSM may not objectively estimate the actual joint angles.

Conclusion

The VSM technique is easy to use and does not require costly equipment. The reproducibility of VSM varies between the different joint/segment angle and between the phases of the gait cycle. When performed by the same observer, in most occasions the VSM technique is reproducible and allows detection of relevant changes after an intervention. The reproducibility of the measurements by different observers is limited. In terms of validity, VSM may differ from 3d-IGA and may not reflect the real sagittal joint position, probably due to the additional movements in the other planes. Therefore, data from VSM must always be interpreted with caution, especially when rotational deviations are present. VSM can be considered as an alternative to 3d-IGA when 3d-IGA is not available or not possible for practical reasons. However, the results and data must always be interpreted in the context of possible measurement errors. 3d-IGA should be preferred whenever possible, particularly in the context of scientific research and in complex clinical questions.

TABLE 1 **Reproducibility of video screen measurement (repeated measurements of the same observer) in six different joint/segment angles during the eight phases of the gait cycle. The table shows the Intraclass Correlation Coefficient (ICC), the Standard Error of Measurements (SEM) and the Smallest Detectable Difference (SDD). The mean expresses the summary of the reproducibility parameters (ICC, SEM, SDD) over all phases of the gait cycle.**

	Shank			Ankle			Knee			Hip			Pelvis			Trunk		
	ICC	SEM	SDD	ICC	SEM	SDD	ICC	SEM	SDD	ICC	SEM	SDD	ICC	SEM	SDD	ICC	SEM	SDD
IC	0.99	0.65	1.80	0.95	2.43	6.75	0.98	1.54	4.26	0.89	2.31	6.41	0.84	1.91	5.30	0.85	5.97	16.55
LR	0.99	5.96	16.51	0.94	1.93	5.35	0.96	1.88	5.19	0.79	2.85	7.91	0.77	1.98	5.50	0.62	3.26	9.05
MST	0.92	1.29	3.59	0.91	3.42	9.48	0.85	3.35	9.30	0.89	2.54	7.04	0.92	1.43	3.98	0.77	2.41	6.68
TST	0.97	1.11	3.07	0.89	3.46	9.58	0.96	2.49	6.89	0.93	2.82	7.83	0.85	2.12	5.88	0.79	2.49	6.9
PSW	0.97	0.97	2.70	0.95	2.27	6.29	0.89	2.74	7.58	0.91	3.29	9.13	0.88	2.11	5.84	0.78	2.36	6.55
ISW	0.96	1.09	3.03	0.97	2.24	6.21	0.94	1.44	3.98	0.95	2.87	7.96	0.92	2.09	5.79	0.84	2.19	6.08
MSW	0.98	1.02	2.82	0.96	2.49	6.91	0.98	1.28	3.55	0.84	2.80	7.77	0.76	2.41	6.69	0.69	2.71	7.51
TSW	0.99	0.81	2.26	0.98	1.68	4.66	0.97	1.91	5.3	0.75	3.02	8.37	0.79	2.09	5.80	0.58	3.23	8.96
Mean	0.97	1.61	4.47	0.94	2.49	6.90	0.94	2.08	5.76	0.87	2.81	7.80	0.84	2.02	5.60	0.74	3.08	8.53

IC = initial contact, LR = Loading response, MST = Mid Stance, TST = Terminal Stance, PSW = Preswing, ISW = Initial Swing, MSW = Mid Swing, TSW = Terminal Swing. ICC = case/(case + occasion + error); SEM = √(occasion + error); SDD = 1.96 x SEM x √2

TABLE 2 **Reproducibility of video screen measurement (measurement of two different observers) in six different joint/segment angles during the eight phases of the gait cycle. The table shows the Intraclass Correlation Coefficient (ICC), the Standard Error of Measurements (SEM) and the Smallest Detectable Difference (SDD). The mean expresses the summary of the reproducibility parameters (ICC, SEM, SDD) over all phases of the gait cycle.**

	Shank			Ankle			Knee			Hip			Pelvis			Trunk		
	ICC	SEM	SDD	ICC	SEM	SDD	ICC	SEM	SDD	ICC	SEM	SDD	ICC	SEM	SDD	ICC	SEM	SDD
IC	0.99	0.70	1.95	0.95	2.60	7.21	0.97	1.87	5.18	0.77	3.12	8.65	0.62	3.06	8.49	0.54	4.76	13.19
LR	0.97	0.95	2.63	0.87	3.47	9.61	0.95	2.07	5.75	0.72	3.31	9.18	0.58	3.19	8.84	0.50	4.52	12.54
MST	0.91	1.50	4.17	0.91	3.28	9.09	0.88	2.87	7.95	0.77	5.32	14.75	0.67	3.06	8.47	0.48	4.26	11.81
TST	0.97	1.13	3.13	0.87	3.73	10.33	0.96	2.55	7.06	0.89	3.30	9.14	0.71	2.82	7.81	0.71	3.12	8.64
PSW	0.95	1.33	3.67	0.88	3.42	9.49	0.89	2.54	7.04	0.87	3.76	10.42	0.79	2.78	7.70	0.68	3.10	8.59
ISW	0.93	1.54	4.28	0.94	2.94	8.14	0.88	1.96	5.42	0.93	3.28	9.11	0.86	2.81	7.78	0.72	3.55	9.85
MSW	0.97	1.17	3.24	0.88	4.02	11.15	0.96	1.70	4.72	0.73	3.84	10.64	0.55	3.94	10.91	0.48	6.78	18.80
TSW	0.98	1.20	3.32	0.96	2.78	7.72	0.96	2.16	5.98	0.67	3.57	9.88	0.60	3.34	9.27	0.50	4.95	13.71
Mean	0.96	1.19	3.30	0.90	3.28	9.09	0.93	2.21	6.14	0.79	3.69	10.22	0.67	3.12	8.66	0.58	4.38	12.14

IC = initial contact, LR = Loading response, MST = Mid Stance, TST = Terminal Stance, PSW = Preswing, ISW = Initial Swing, MSW = Mid Swing, TSW = Terminal Swing. ICC = case/(case + occasion + investigator + error); SEM = √occasion + investigator + (occasion x investigator); SDD=1.96 x SEM x √2

FIGURE 1 **Video Screen measurement of 6 sagittal joint/segment angles on video screen. With the aid of a custom made, open-source software package (the Moxie Viewer®, www.smalll.nl) angular measurements for the shank, ankle, knee, hip, pelvic tilt and trunk tilt were performed in the sagittal plane using a multi-goniometer. The software allows to synchronously observe gait parameters and video recordings. The speed of the videos can be freely selected, including slow motion and freeze frame techniques.**

TABLE 3 **Agreement of Video Screen Measurement and 3d instrumental gait analysis expressed by the 95% limits of agreement (LoA = absolute difference +/- 2 SD). There is a large variation between the measurements for all angles.**

	95% LoA (Degrees)
Peak Ankle DFL Stance	5.7 +/–15.2
Peak Knee EXT Stance	0.4 +/–13.4
Peak Hip EXT Stance	1.4 +/–15.6
Peak Ankle DFL Swing	12.0 +/–14.6
Peak Knee FLX Swing	1.4 +/–11.4
Peak Hip FLX Swing	2.6 +/–18.8

DFL = Dorsiflexion, EXT = Extension, FLX = Flexion

FIGURES 2a–2f **Bland-Altman plots expressing the agreement of three-dimensional instrumented gait analysis andvideo screen measurement for three different joint angles (Fig. 2a: Ankle angle in the stance phase, Fig. 2b: Ankle angle in the swing phase, Fig.2c: Knee angle in the stance phase, Fig. 2d: Knee angle in the swing phase, Fig. 2e: Hip angle in the stance phase, Fig. 2f: Hip angle in the swing phase). The y-axes represent the difference between video screen measurements and three-dimensional measurements, the x-axes represents the mean of video screen measurements and three dimensional measurements. The straight line corresponds to the mean difference between the measurements; the dotted lines correspond to two standard deviations. There is a large variability between the measurements for all angles. For the ankle angles (Fig. 2a and Fig. 2b) the video screen measurements systematically show less plantar flexion than the three dimensional measurements.**

Fig. 2a

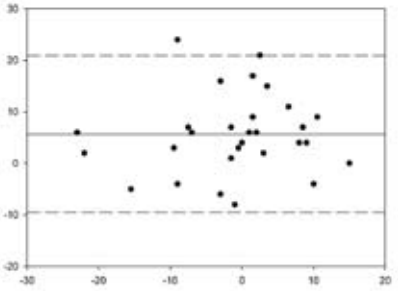


Fig. 2b

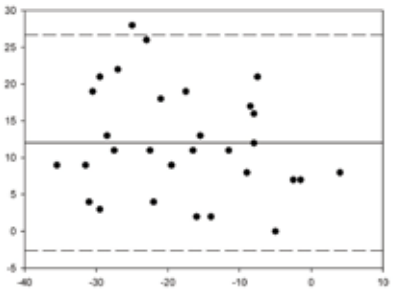


Fig. 2c

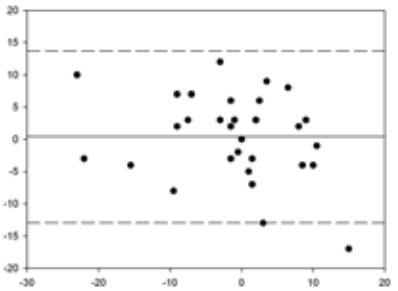


Fig. 2d

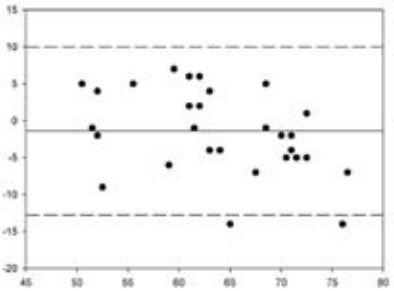


Fig. 2e

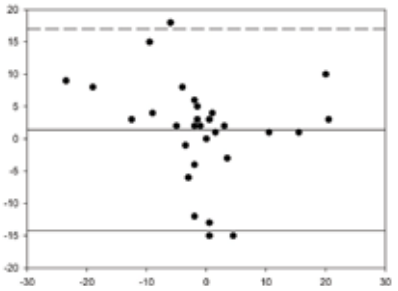
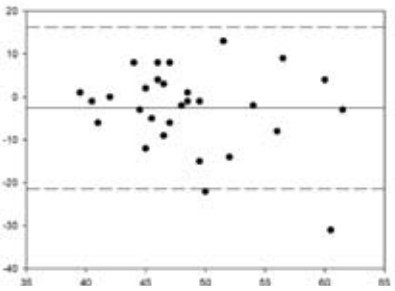


Fig. 2f



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Chapter 6

Effect of Selective Dorsal Rhizotomy on Gait in Children with Bilateral Spastic Paresis: Kinematic and EMG-Pattern Changes

Sebastian Grunt, Wouter J. P. Henneman, Mark J. Bakker, Jaap Harlaar, Willem J. R. van Ouwerkerk, Jules G. Becher, Petra E.M. van Schie, Alexander Reeuwijk and R. Jeroen Vermeulen

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Abstract

Introduction

Selective dorsal rhizotomy (SDR) is an effective treatment for reducing spasticity and improving gait in children with spastic cerebral palsy. Data concerning muscle activity changes after SDR treatment are limited.

Patients and methods

In 30 children who underwent SDR a gait analysis was performed before and 12 to 24 months postoperatively. Subjects walked on a 10-m walkway at comfortable walking speed. Biplanar video was registered and surface EMG was recorded. Sagittal knee angles were measured from video and observational gait assessments were performed using the Edinburgh Gait Assessment Scale (EGAS).

Results

The EGAS significantly improved after SDR ($p<0.001$). There were significant improvements of the knee angle kinematics ($p<0.001$). Only slight changes in EMG activity were observed. The activity of the m. gastrocnemius (GM) decreased and a late peak appeared in stance, the activity of the m. semitendinosus (ST) increased in stance. The activity of the m. rectus femoris (RF) decreased in swing.

Conclusion

SDR improved overall gait performance but EMG changes were only slight. Better timing of the GM in stance and reduced activity of RF in swing may have increased knee flexion in swing. Reduced hamstrings spasticity may have led to postural instability in the hip.

Introduction

Cerebral palsy (CP) describes a group of permanent, activity-limiting disorders of movement and posture development caused by non-progressive disturbances in the developing fetal or infant brain [25]. Its most common form is spastic CP, which can be classified as either unilateral or bilateral [31]. Spasticity is defined as a velocity-dependent increase of the tonic stretch that results from a reduction of lower motor neurons' inhibitory impulses due to an upper motor neuron lesion [26]. Spasticity can cause pain, muscle shortening, and orthopaedic malformations, and it can severely interfere with functional abilities and gait pattern.

Different treatment modalities have been proposed to reduce spasticity in children with spastic CP (29). Selective dorsal rhizotomy (SDR) is a neurosurgical intervention aimed at reducing spasticity in the legs. By partially transsecting the posterior lumbosacral rootlets, SDR reduces the excitatory sensory input from the legs that enters the spinal cord at the dorsal root. SDR has mainly been studied in patients with bilateral spastic CP. Various studies have shown that SDR reduces spasticity [16–19, 27, 28, 34, 39], improves passive range of motion in the lower extremities [6, 21, 28], improves gross motor function [5, 15, 17–19, 21, 27, 28, 35] and has a positive influence on the performance of activities of daily life [5, 20, 21, 35]. The selection criteria for SDR treatment differ between centres; however, SDR is mostly performed in patients who have an involvement of multiple muscle groups in the lower legs, in which spasticity is the only – or at least the predominant – form of motor disturbance and in which spasticity negatively influences gross motor function. Furthermore, the goal of SDR depends on the severity of the motor impairment. When SDR is performed in patients with preserved walking ability, the main goal is to improve the gait pattern and the gait efficiency.

Various studies have assessed gait performance in children spastic with CP who underwent SDR [1–8, 14, 28, 30, 32–37]. In most studies, time-distance parameters (such as gait velocity, cadence and stride length) and the kinematics of different joints (mainly in the sagittal plane) have been assessed, using three-dimensional [1, 3, 5, 6, 8, 32, 33] or two-dimensional instrumented gait analysis [4, 14, 30, 36, 37]. It has been shown that SDR provides short-term improvements in the gait kinematics of the hip, knee and ankle and has a positive influence on the foot-floor contact pattern [1–6, 8, 32, 33, 36]. SDR's influence on time-distance parameters was inconsistent among the studies [1, 3, 4, 8, 32, 33, 36]. Langerak et al. have shown that the improvement in the hip and knee kinematics persist over the long term (20 years postoperatively) [14]. Three studies have shown significant improvements in overall gait performance after SDR [5, 34, 35]. Trost et al. used the Gillete Gait Scale (based on 3-dimensional instrumental gait analysis) and examined a larger cohort of 136 patients with CP undergoing SDR [34]; Chan et al.

used the Observational Gait Scale (based on observational gait analysis) in a group of 21 children undergoing SDR [5] and our group used the Edinburgh Gait Assessment Scale (EGAS, based on observational gait analysis) to describe the gait patterns after SDR in the first 9 patients who underwent SDR in our centre [35]. In studies by Trost et al. and Chan et al., gait efficiency was also examined by measuring oxygen consumption during gait. Whereas Trost el al. at found a significant improvement in gait efficiency [34], Chan et al. only documented the results of oxygen consumption after SDR in the patients with the lowest grade of motor impairment and could not find a significant change [5].

The gait pattern of children with spastic CP is influenced by a loss of voluntary motor control and the use of primitive movement patterns. Surface-EMG gait recordings often show a co-activation pattern and/or prolonged and out-of-phase muscle activation, which results from an increased stretch response. It could be hypothesised that by reducing spasticity, SDR would have an influence on these EMG patterns. Five studies used surface EMG measurements to assess the changes in lower extremity muscle activity after SDR [1, 3, 4, 7, 32]. Whereas most of these studies used a qualitative assessment for the EMG analysis and provided their results in a descriptive way [3, 4, 7, 32], Abel et al. quantitatively analysed the EMG patterns before and after SDR [1]. In summary, the EMG patterns remained unchanged in most cases. However, Abel et al. found a significant decrease in the activity of the m. rectus femoris in the early swing phase and a significant decrease in the activity of the m. gastrocnemius in the loading response [1]. They concluded that the stretch response influences gait in patients with CP and that SDR attenuates the stretch response and consequently affects gait mechanics.

Although some studies extensively describe the selection criteria patients must meet to qualify for SDR, to our knowledge, the influence of preoperative characteristics, such as the severity of motor impairment or the severity of preoperative gait abnormalities, on outcome has not been well documented. However, because SDR is irreversible, it is important to select the most ideal candidates for the procedure. We therefore aimed to assess outcomes, as determined by overall gait performance, in all patients who underwent SDR in our institution between January 1998 and December 2009. We aimed to determine if there were associations between the preoperative patient characteristics and the improvement of gait abnormalities after SDR, to describe the influence of SDR on EMG patterns using a qualitative and a quantitative method, and to describe the changes in knee joint kinematics after SDR.

Methods

Participants

In the present study, we included patients who underwent SDR at the lumbar level at the VU University Medical Center, Amsterdam, the Netherlands, between August 1998 and March 2009 and for whom we could obtain a gait analysis before SDR and 12 to 24 months postoperatively. The VU Medical Center's eligibility criteria for SDR are as follows: 1. Bilateral spasticity of the lower extremities that interferes with walking performance; 2. Spasticity (defined as velocity-dependent resistance to passive stretch) in at least 6 muscle groups of the lower limbs; 3. Sufficient force in the quadriceps femoris muscle (ability to squat at least seven times) and the hip extensors (ability to kneel with extended hips, with support allowed for balance); 4. Absence of structural orthopaedic deformities or contractures at the hip, knee or ankle; 5. Moderate to good selective motor control in the lower limbs; 6. Gross Motor Function Classification System (GMFCS) [23] Level I, II or III; 7. Good support from parents and the rehabilitation setting. As we have shown that SDR improves gross motor function in patients with both normal MRIs and periventricular leucomalacia (PVL) in neuroimaging [9], a strict diagnosis of cerebral palsy (defined as non-progressive disturbances that occurred in the developing fetal or infant brain) [25] was not a criterion for inclusion in our study. However, the majority of the patients included did show evidence of PVL in neuroimaging.

Operative Procedure and Rehabilitation

The same neurosurgeon (WVO) performed the SDR in all children. After laminotomy of L2 to L5 and opening of the dura, the dorsal roots L2 to S1 were exposed and separated into different rootlets. Transsection of the rootlets was performed after electrostimulation, according to palpable muscle contraction and EMG response. No more than 50% of the rootlets were transsected at each level. To prevent sexual and bladder disturbances, rootlets/fascicles showing electrical responses upon stimulation of the penis/clitoris were spared. Post-operative rehabilitation included intensive physical therapy for 12 months.

Gait Analysis

Gait analysis was performed before SDR and 12 to 24 months after SDR. The subjects walked barefoot along a 10-m walkway at a self-selected, comfortable speed. Video recordings of both legs were taken in the sagittal and frontal plane. A custom-made, open-source software package that allows the researchers to simultaneously observe gait parameters and video recordings (the MoXie Viewer®, www.smalll.nl) was used for the data analysis. This software package allows users to freely select the video speed, including slow motion and freeze-frame techniques. The software includes a graphical multi-goniometer that allows the measurement and display of

six sagittal angles. The repeatability and validity of this method has been described previously [10]. In the present study, we measured only the sagittal knee angle kinematics (the minimal knee extension in swing, maximal knee flexion in stance and the total knee excursion). The measurements were performed by the same observer for three representative strides, and the mean of these three measurements was used for the statistical analysis.

The main outcome parameter in the present study was the overall gait performance after SDR. For this purpose, we used the EGAS, which is an observational gait analysis scale that assesses a patient's gait deviation with 17 different items on an ordinal scale and examines the movements of various joints/segments (the foot/ankle, the knee, the hip, the pelvis and the trunk) in the sagittal and the frontal plane separately for the stance and the swing phase [24]. A normal gait pattern would receive an EGAS score of 0, and maximal gait deviation would be scored as 34. Good intra- and inter-observer reliability have been reported for the EGAS [22, 24] and it has been shown to objectively represent the severity of gait deviation in patients with CP [11].

Surface EMG was registered for the m. rectus femoris (RF), the m. vastus lateralis (VL), the m. gastrocnemius medialis (GM), the m. tibialis anterior (TA) and the m. semitendinosus (ST). Raw EMG signals were high-pass filtered (20 Hz, 2nd order Butterworth) to remove movement artefacts and rectified and smoothed at 2 Hz to obtain the envelope. Time-normalisation (0 to 100% of the gait cycle) of the EMG envelope from at least three strides was performed and averaged using custom-made software (MatLab 6.0®, Mathworks, Natick, Massachusetts, USA, www.mathworks.com) to obtain the EMG profile. The amplitude of the EMG envelope was normalised to 100% of maximal EMG activity. To assess the amount of abnormal activity in the EMG signal, the EMG measurements were compared with normal EMG profiles [38] in the following manner: First, the gain of the original EMG signal was matched with the optimal match method, according to Hof et al. [12]. Next, the mean difference between the gain optimised measurement and normal EMG activity was calculated and provided as a percentage. Finally, for each muscle, separate qualitative evaluations of the EMG profiles were performed for the stance and swing phase, using explicit criteria based on patterns associated with spasticity or co-contraction (as shown in Table 1) recognised at two separate time points by two different observers.

TABLE 1 **Criteria for the qualitative EMG assessment. For each muscle, improvement and deterioration were defined separately for the stance and the swing phase according to these criteria.**

	Stance		Swing	
	Improvement	**Deterioration**	**Improvement**	**Deterioration**
RF	More distinct phased activity Longer activity at initial contact	Increased continuous activity Decreased peak in loading response	Decreased activity in comparison to stance Decreased activity in the total swing phase	
VL	More distinguishable peak activity Decrease in continuous activity	Prolonged activity Increased continuous activity	Decreased activity at initial swing and mid-swing	Increased relative activity
ST	Disappearance of peak activity in terminal stance	Appearance of a peak in terminal stance Peak after loading response Increased continuous activity Flattening of the EMG signal	Decreasing activity in initial swing and mid-swing Appearance of a peak in terminal swing Increase of peak/lowest point ratio	Lack of a peak in terminal swing Increased activity in initial swing and mid-swing Flattening of the EMG signal
GC	Decreased activity at the start of stance Appearance of a peak at terminal stance	Increased activity in the stance phase Disappearance of peak at terminal stance	Decline of activity, with lowest point at mid-swing Disappearance of rising near the end of swing	Increased activity in terminal swing Increased continuous activity
TA	Appearance of peak activity in loading response Decreased activity in mid-stance and terminal stance	Prolonged continuous activity Increased activity in mid-stance and terminal stance	Decrease of the peak in mid-swing Rising activity near the end of swing	Decreased activity in comparison to stance Decline near the end of swing Increased continuous activity

Abbreviations used: RF = M. rectus femoris, VL = M. vastus lateralis, ST = M. semitendinosus, GC = M. gastrocnemicus, TA = M. tibialis anterior

Statistical Analysis

Instead of reporting the results separately for the left and the right leg, the assessments of both legs were pooled for the statistical analysis. To compare the scores for single EGAS items and the total EGAS score before and after SDR, we used the nonparametric Wilcoxon test. The change in the EGAS total score after SDR was correlated with the following parameters, using Spearman's rank correlation quotient: Age at SDR, Gross Motor Function Classification Score [23] before SDR and EGAS total score before the SDR. The kinematic measurements of the knee joint before and after SDR were compared with a Student's t-test. For the qualitative EMG evaluation the changes in the EMG signals have been reported in a descriptive way. The intra-observer and the inter-observer reliability of the qualitative EMG evaluation has been calculated by Kappa statistics. Agreement strengths for the Kappa values were classified according to Landis and Koch [13]. To compare the amount of abnormal EMG activity before and after SDR, the difference between the optimised measurement and normal EMG activity was compared with the nonparametric Wilcoxon test. For the statistical analysis, we used SPSS® version 15.0 for Windows. The level of significance was set at 0.05.

Results

Participants

Between January 1998 and March 2009, 44 patients with bilateral spasticity and preserved walking ability underwent SDR at the VU University Medical Center, Amsterdam, the Netherlands. In 30 patients (mean age: 6.54 years, SD: 1.96 years; range: 2.75 years to 13.16 years; 19 boys, 11 girls) a gait analysis was performed preoperatively and 12 to 24 months postoperatively (mean follow-up duration: 14.97 months; SD: 2.64 months; range: 12 to 21 months). Before the SDR, 3 patients (10%) were at GMFCS Level I, 9 patients (39%) were at GMFCS Level II, and 18 patients (60%) were at GMFCS Level III. Of the 18 patients at GMFCS Level III, 12 used a posterior walker, 5 used an anterior walker and one walked with caregiver assistance. After the SDR, all patients who walked independently before SDR (GMFCS Level I or II) did not use an assistive device. One of the patients who walked with a posterior walker before SDR walked independently after SDR, and another patient who walked with a posterior walker before SDR walked with three-point crutches after SDR. The remaining 10 patients were still walking with a posterior walker. The 5 patients who walked with an anterior walker before the SDR walked with a posterior walker after the SDR, and the patient who walked with caregiver assistance before SDR still required caregiver assistance after the SDR.

Overall Gait Assessment and Correlation with Preoperative Parameters

In Table 2, EGAS results before and after the SDR are listed separately for independent ambulators (GMFCS I and II) and for patients who walked with assistive devices before SDR (GMFCS III). Figure 1 provides boxplots for the EGAS total scores before and after SDR for the two subgroups. In summary, there were significant improvements in the overall gait performance for all patients ($p<0.001$). The improvement in the EGAS scores was significant in both subgroups: the independent ambulators ($p<0.001$) and the patients who walked with assistive devices before SDR ($p=0.002$). However, the improvements were much more pronounced among the independent ambulators. The improvements after SDR correlated significantly with the preoperative GMFCS level (rho = –0.413, $p=0.001$), but there was no significant correlation between the patient's age when SDR was performed and the outcome after SDR (rho = 0.207, $p=0.112$), nor between the preoperative EGAS score and the improvement in the overall gait performance after the SDR (rho = 0.223, $p = 0.086$). Table 3 illustrates the changes after SDR in the single EGAS items. Significant improvements were found mainly in the foot and ankle, namely the heel lift ($p<0.001$), the maximal ankle dorsiflexion in stance ($p<0.001$), the foot rotation ($p=0.037$), the foot clearance in swing ($p<0.001$), and the maximal ankle dorsiflexion in swing ($p<0.001$). Furthermore, there was significant improvement in the knee progression angle ($p=0.030$), the knee flexion in terminal swing ($p=0.002$) and the peak hip extension in stance ($p=0.014$).

TABLE 2 **Total Score of the Edinburgh Gait Assessment Scale before and after SDR. Data for both legs have been pooled. The results are listed separately for children who walked without assistive devices before the SDR (GMFCS I and II = independent ambulators), for children who walked with assistive devices before SDR (GMFCS III = assisted ambulators) and for all children in the study (GMFCS I through III).**

		Before SDR	After SDR	Difference	
	n	Mean (SD)	Mean (SD)	Mean (SD)	p-Value
Independent ambulators	24	15.6 (3.7)	8.3 (3.4)	7.3 (3.1)	<0.001
Assisted ambulators	36	18.2 (3.8)	15.4 (4.0)	2.8 (5.0)	0.002
All subjects	60	17.2 (3.9)*	12.6 (5.1)**	4.6 (4.8)***	<0.001

***Significant correlation with preoperative GMFCS (rho = 0.375, p = 0.003)**
**** Significant correlation with preoperative GMFCS (rho = 0.689, p < 0.001)**
*****Significant correlation with preoperative GMFCS (rho = 0.413, p = 0.001)**

FIGURE 1 **Total Score of the EGAS before and after SDR**

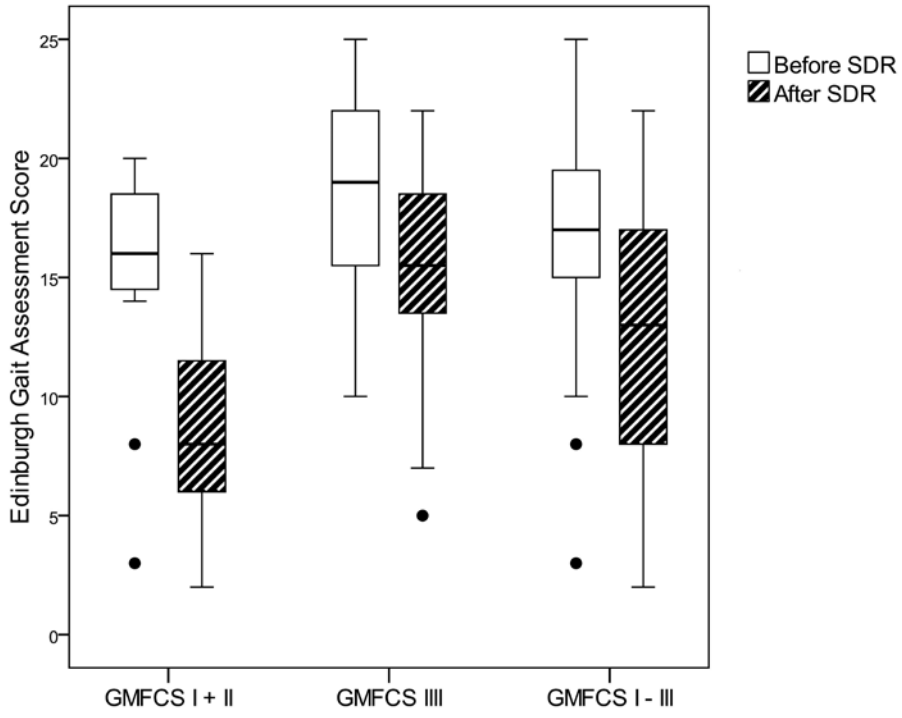


TABLE 3 **Single items of the Edinburgh Gait Assessment Scale before and after SD**

	Mean (SD)	STANCE Mean (SD)	p-value		Mean (SD)	SWING Mean (SD)	p-value
Ankle							
	Before SDR	After SDR			Before SDR	After SDR	
Initial contact	1.90 (0.35)	1.42 (0.56)	<0.001	Clearance swing	0.82 (0.50)	0.35 (0.48)	<0.001
Heel lift	1.83 (0.46)	1.03 (0.58)	<0.001	Max. ankle dorsiflexion	1.50 (0.68)	0.67 (0.66)	<0.001
Max. ankle dorsiflexion	1.52 (0.62)	0.78 (0.67)	<0.001				
Hind foot valgus/varus	0.52 (0.50)	0.68 (0.68)	NS				
Foot rotation	0.63 (0.52)	0.42 (0.56)	0.037				
Knee							
	Before SDR	After SDR			Before SDR	After SDR	
Knee progression angle	0.75 (0.63)	0.52 (0.50)	0.030	Terminal swing	1.80 (0.48)	1.53 (0.65)	0.002
Peak extension stance	1.23 (0.81)	1.05 (0.81)	NS	Peak flexion swing	0.68 (0.75)	0.88 (0.82)	NS
Hip							
	Before SDR	After SDR			Before SDR	After SDR	
Peak extension stance	1.10 (0.86)	0.78 (0.88)	0.014	Peak flexion swing	0.95 (0.67)	0.93 (0.69)	NS
Pelvis							
	Before SDR	After SDR					
Obliquity mid-stance	0.40 (0.56)	0.30 (0.46)	NS				
Rotation mid-stance	0.40 (0.56)	0.25 (0.44)	NS				
Trunk							
	Before SDR	After SDR					
Peak sagittal position	0.78 (0.80)	0.65 (0.76)	NS				
Max. lateral shift	0.35 (0.51)	0.33 (0.51)	NS				

Knee Angle Kinematics

The results of the knee joint kinematics are summarised in Table 4. In summary, there was a significant improvement in knee angle kinematics after SDR. The maximal knee extension in the stance phase improved from 19.6 degrees (SD 14.6) before SDR to 11.4 degrees (SD 13.6) after SDR ($p < 0.001$); the maximal knee flexion in swing improved from 61 degrees (SD 8.9) before SDR to 66.5 degrees (SD 10.5) after SDR ($p < 0.001$); and the total knee joint excursion improved from 41 degrees (SD 14.7) before SDR to 59.8 degrees (SD 15.7) after SDR.

TABLE 4 **Knee angle kinematics before and after SDR**

	GMFCS I-III			GMFCS I & II			GMFCS III		
	Before SDR	After SDR	p	Before SDR	After SDR	p	Before SDR	After SDR	p
Peak knee angle stance (SD)	19.6° (14.6)	11.4° (13.6)	<0.001	13.6° (10.5)	7.9° (7.2)	0.003	23.9° (15.8)	14.0° (16.4)	<0.001
Peak knee angle swing (SD)	61.0° (8.9)	66.5° (10.5)	<0.001	57.5° (9.2)	62.2° (9.3)	0.004	63.5° (8.0)	69.4° (10.5)	0.002
Knee excursion (SD)	41.0° (14.8)	59.8° (15.7)	<0.001	43.9° (11.5)	61.2° (10.5)	<0.001	38.9° (16.6)	58.8° (18.7)	<0.001

Surface EMG

Because the type of assistive device used might influence the EMG pattern, only the patients who walked with the same assistive device before and after the SDR (12 independent ambulators, 10 patients who walked with a posterior walker and 1 patient who walked with caregiver assistance) were included in the EMG analysis.

The results of the qualitative EMG measurements are summarised in Table 5a, which also provides the Kappa values for both intrarater and the interrater reliability. In summary, the intrarater reliability was better than the interrater reliability, and the reliability was better for the swing phase than for the stance phase. The intrarater reliability was perfect for the GM in the swing phase (Kappa = 1.0) and almost perfect for the ST in the swing phase (Kappa = 0.922); substantial for the TA in the stance phase (Kappa = 0.750), the ST in the stance phase (Kappa = 0.698), the RF in the swing phase (Kappa = 0.651), the TA in the swing phase (Kappa = 0.625) and the GM in the stance phase (Kappa = 0.606); and moderate for the VL in the stance phase (Kappa = 0.525) and the RF in the stance phase (Kappa = 0.479). The interrater reliability was substantial for the RF in the swing phase (Kappa = 0.721) and the VL in the swing phase (Kappa = 0.776); moderate for the VL in the stance phase (Kappa = 0.495, the TA in the stance phase (Kappa = 0.481) and the TA in the swing phase

(Kappa = 0.455); and fair for the RF in the stance phase (Kappa = 0.361), the GM in the stance phase (Kappa = 0.398) and the ST in the swing phase (Kappa = 0.232). Kappa statistics could not be calculated for the interrater reliability of the ST in the stance phase and the GM in the swing phase because only one of the two observers graded both, improvements and deteriorations on the two-point ordinal scale.

In the stance phase, the main changes were observed in the GM, which showed decreased activity in the stance phase and/or the appearance of a peak in terminal stance in 58% of the patients, and in the ST, which showed deterioration with increased continuous activity in 64% of the patients. In the swing phase, the main changes were found for the RF, which showed decreasing activity when compared with stance in 56% of the patients. Table 5b summarises the results of the quantitative EMG analysis separately for all patients (GMFCS I to III), the subgroups of independent ambulators (GMFCS I & II) and the patients who used an assisted device before the SDR (GMFCS III). There was significant improvement, with more normal activity of the TA anterior ($p = 0.010$) and a significant deterioration with less normal activity of the ST ($p < 0.001$). More normal TA activity was also found in the subgroup of patients that walked without an assistive device ($p = 0.001$); however, the subgroup of patients that used an assistive device did not show significant improvement of the TA after SDR.

Discussion

In the present study, we describe the outcomes for a group of patients with bilateral spasticity that underwent SDR at the VU University Medical Center (Amsterdam, the Netherlands) with respect to their gait performance 12 to 24 months after surgery. Different outcome assessments were used to describe their improvements after SDR.

The main outcome parameter was the EGAS, a valid and reliable measurement for the assessment of gait abnormalities in patients with CP that describes the overall gait performance [22, 24]. Consistent with previous studies [5, 34, 35], we found a significant improvement in the overall gait performance after SDR. Similar to previous studies [1–5, 8, 32, 33], we found significant improvements, mainly in the foot and ankle. The improvement was better in patients who walked without an assistive device before SDR, and there was a clear correlation between the improvement in overall gait performance after SDR and the preoperative GMFCS level.

Similar to the present study, most studies that assessed gait performance after SDR included patients at GMFCS Levels I to III [1–5, 14, 30, 34, 36, 37]. Trost et al. used multidimensional assessment methods, including 3-dimensional gait analysis and oxygen consumption measurement, to examine a cohort of 136 patients after SDR [34]. The only outcome parameter that differed between patients with different levels of motor impairment was the improvement of gait speed after SDR, which was more pronounced in the patients with GMFCS Level III (possibly due to a ceiling effect in GMFCS Levels I and II). No other study has compared the outcome of SDR in terms of gait performance in patients with differing levels of impairment. As a rule, patients with weak muscle strength and poor selective muscle control are not considered good candidates for an SDR because spasticity may support the gait of children with CP who are weak or have lost voluntary motor control. In the present study, the eligibility criteria for SDR included good strength in the quadriceps femoris and the hip-extensors and good selective muscle control. However, strength in the plantar flexors of the ankle was not an inclusion criterion. We therefore assume that muscle weakness, especially in the plantar flexors of the ankle, might have been more pronounced in the patients with GMFCS Level III and might explain the reduced improvement compared with patients at GMFCS Levels I and II.

Additionally surface EMG measurements were used to assess changes in muscle activity during gait before and after SDR. The EMG data were analysed quantitatively and qualitatively. Most previous studies assessed changes in surface EMG patterns only with a qualitative method [3, 4, 7, 32]. Only one study used a quantitative method to assess changes in surface EMG measurements after SDR [1]. Both methods have advantages and disadvantages. Qualitative analysis allows observers to define the changes activity patterns, such as the timing of the muscle activity during the gait cycle, for a single patient, though the method is observer-dependent. Previous studies have not reported reliability measures for the qualitative EMG analysis [3, 4, 7, 32]. In the present study, the intrarater reliability varied from moderate to perfect, and the interrater reliability varied from fair to substantial, which we consider acceptable in a clinical setting. The quantitative method represents a more objective method; however, it only allows a description of general changes. Thus no conclusions about changes in the timing of the EMG activity in a single patient can be drawn.

In contrast with the significant changes in the overall gait performance but consistent with previous findings [1, 3, 4, 7, 32], the changes in EMG activity after SDR were minimal. The most striking changes have been found for the ST, which showed significantly less normal activity in the quantitative analysis and prolonged activity in the qualitative analysis of the stance phase in the majority of the patients. The hamstrings can act as knee flexors and as hip extensors. In the stance phase, the hamstrings have a stabilizing function in the hip joint and the pelvis. We assume that the deterioration of activity in the ST in the stance phase might be a compensatory mechanism resulting from of a change in postural stability after SDR. Consistent with Abel et al. [1], we also observed a decreasing RF activity during the swing phase in the qualitative EMG analysis.

Furthermore, sagittal knee angle kinematics were measured before and after SDR. We found a significant improvement in the maximal knee extension in the stance phase. In the stance phase, the knee extension is coupled with plantar flexion in the ankle at the end of the stance phase, resulting from the push-off activity of the GM. In the present study, we found an improvement in GM activity in the majority of the patients, with the appearance of a peak at terminal stance, especially in patients who walked without an assistive device. In addition, we found an improved knee flexion in the swing phase and an improvement in overall knee joint excursion. Knee flexion in the swing phase can be reduced by the decreased push-off activity of the GM at the end of the stance phase, by persistent RF activity in the swing phase, or both. Consistent with the EMG findings described above, we assume that the improvement of knee flexion in the swing phase might be caused by a decreased RF stretch response after SDR during the fast knee flexion movement in the beginning of the swing phase and a more pronounced GM peak activity at the end of the stance phase, leading to better push-off activity.

Conclusions

We conclude that SDR leads to improvements in the overall gait performance, mainly in patients who walk without assistive devices, and improvements are observed in the foot and ankle as well as in the knee angle kinematics. Despite these improvements, there are only slight changes in EMG activity after SDR. Better GM push-off activity might explain the knee extension improvement in the stance phase. Reduced EMG activity in the RF might permit increased knee flexion in swing, and a deterioration of the hamstrings activity might be caused by decreased postural stability.

Limitations

The main outcome parameter in the present study was the EGAS, which is a reliable and valid measurement tool. In addition, we performed kinematic measurement of the knee angle on a video screen with the aid of a custom-made software program. However, the gold standard for the evaluation of gait abnormalities in children with CP is three-dimensional gait analysis. The measurement tool used in the present study has been evaluated for its reliability and validity, and the changes that we detected in the knee angles were mostly within the limits of measurement errors [10]. However, because the measurement tool is limited to two dimensions, it may not reflect the real sagittal joint position; furthermore, it does not permit the measurement of kinematic changes in planes other than the sagittal. Additional movements in the other planes cannot be assessed, and the kinematic data provided in this study must be interpreted with caution.

TABLE 5a **Results of the qualitative EMG analysis The Kappa values were calculated as follows: Kappaintra = Agreement of Observer 1's measurements; Kappainter = Agreement between the measurements of Observer 1 and Observer 2**

		Stance					Swing				
	Pairs	**Improvement**	**Deterioration**	**No change**	**Kappa$_{intra}$**	**Kappa$_{inter}$**	**Improvement**	**Deterioration**	**No change**	**Kappa$_{intra}$**	**Kappa$_{inter}$**
RF	32	4 (12 %)	8 (25 %)	20 (63 %)	0.479	0.361	18 (56 %)	3 (9 %)	11 (35 %)	0.651	0.721
VL	29	4 (14 %)	7 (24 %)	18 (62 %)	0.525	0.495	4 (14 %)	0 (0 %)	25 (86 %)	(95%)*	0.776
GM	26	15 (58 %)	6 (23 %)	5 (19 %)	0.606	0.398	1 (%)	0 (0 %)	25 (%)	1.000	(90%)*
TA	23	8 (35 %)	7 (30 %)	8 (35 %)	0.750	0.481	1 (4 %)	2 (8 %)	20 (78 %)	0.625	0.455
ST	33	4 (12 %)	21 (64 %)	8 (24 %)	0.698	(66%)*	6 (18 %)	6 (18 %)	21 (64%)	0.922	0.232

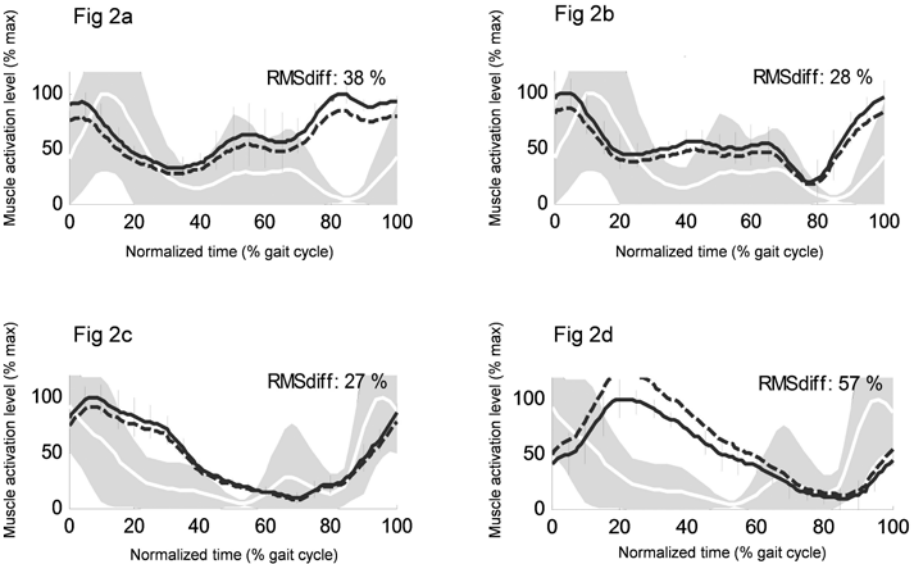
***Calculation of Kappa value was not possible because one observer scored the whole range of changes, whereas the other did not. The value in parentheses represents the percentage of agreement. Abbreviations used: RF = M. rectus femoris, VL = M. vastus lateralis , GM = M. gastrocnemius medialis, TA = M. tibialis anterior, ST = M. Semitendinosus**

TABLE 5b **Results of the quantitative EMG analysis. The mean values represent the mean difference between the gain-optimised measurement and normal EMG activity.**

	GMFCS I–III				**GMFCS I – II**				**GMFCS III**			
		Before SDR	**After SDR**			**Before SDR**	**After SDR**			**Before SDR**	**After SDR**	
	N	Mean (SD)	Mean (SD)	P Value	N	Mean (SD)	Mean SD	P Value	N	Mean (SD)	Mean (SD)	P Value
RF	32	37.1 (8.6)	35.2 (8.8)	0.141	18	37.1 (8.6)	36.3 (10.5)	0.371	14	37.0 (8.9)	33.7 (6.2)	0.208
VL	29	29.6 (7.5)	32.2 (6.8)	0.051	19	26.3 (6.1)	29.2 (4.6)	0.131	10	35.8 (6.1)	37.9 (7.0)	0.153
GM	26	22.4 (5.2)	20.9 (5.6)	0.115	18	22.2 (5.6)	19.7 (3.9)	0.083	8	22.9 (4.6)	23.7 (7.8)	0.735
TA	23	30.8 (7.2)	26.3 (7.3)	0.010	17	30.4 (7.7)	23.2 (3.3)	0.001	6	32.0 (6.2)	35.0 (8.8)	0.462
ST	33	32.2 (8.8)	46.0 (11.4)	<0.001	19	33.3 (9.7)	43.8 (11.3)	0.007	14	30.8 (7.4)	49.0 (11.2)	0.001

Abbreviations used: RF = M. rectus femoris, VL = M. vastus lateralis , GM = M. gastrocnemius medialis, TA = M. tibialis anterior, ST = M. semitendinosus

FIGURES 2a–2d **Typical example of EMG measurement. The figures show a typical example of EMG measurements before SDR in the M. rectus femoris (Fig 2a) and the M. semitendinosus (Fig 2c) and after SDR in the M. rectus femoris (Fig 2b) and the M. semitendinosus (Fig 2d). The x-axis represents the gait cycle in percent and the y-axes represents the relative amplitude of the surface EMG signal in % peak value. The dark line represents the mean of the original EMG measurements and the vertical dark bars represent one standard deviation. The dotted dark line represents the matched gain according to the method of Hof et al. (12). The grey area shows the spread of normal EMG profiles according to the data of Winter et al. (38), the white line represents the mean normal profile. RMSdiff represents the mean difference between the gain optimized measurement and the normal values in percent. Note the prolonged and increased activity in the M. semitendinosus in the stance phase (Fig 2c and 2d) and the decreasing activity of the M. rectus femoris in the swing phase (Fig 2a and 2b) after SDR.**



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Chapter 7

Long-term outcome and adverse effects of selective dorsal rhizotomy in children with cerebral palsy – a systematic review

Sebastian Grunt, Jules G. Becher and R. Jeroen Vermeulen

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Abstract

Aim

To assess the long-term outcome and adverse events of selective dorsal rhizotomy (SDR) in children with spastic cerebral palsy (CP).

Method

Studies were selected based on the following inclusion criteria: Children with CP that underwent SDR with a follow-up period of at least five years. The following databases were searched: MEDLINE, Web of Science, Embase, PEDro, and the Cochrane library. Studies meeting the inclusion criteria were scored by two reviewers, who graded the level of evidence and the quality/conduct of the studies. Outcomes were classified according to the International Classification of Functioning, Disability and Health (ICF).

Results

Only three of the twenty-one studies who met the inclusion criteria allowed a tentative conclusion on outcome. There is moderate evidence that SDR has a positive long-term influence on the ICF body structure and body function domains but there is no evidence that SDR has an influence on the ICF activity and participation domains. Spinal abnormalities seem to be common, but no conclusion can be drawn about their relation to SDR.

Interpretation

There is lack of evidence concerning the long-term outcomes after SDR. Future studies need to clarify the long-term influence of SDR – especially in the ICF domains of activity and participation.

Selective Dorsal Rhizotomy (SDR) is a neurosurgical procedure that is mainly performed at the lumbar level to reduce spasticity in patients with bilateral spastic paresis. The procedure is performed mostly in children with spastic cerebral palsy. Spasticity is defined as a velocity-dependent increase in the tonic stretch response with excessive tendon jerk reflexes, and it is caused by the reduction of inhibitory impulses on lower motor neurons.[1] SDR reduces the excitatory input to the spinal cord from the lower limbs with incomplete trans-section of the (sensory) posterior lumbosacral rootlets. The outcomes after SDR are the subject of various studies, which were summarised in 2001 in a systematic review.[2] Furthermore, in 2002 a meta-analysis of three randomised studies comparing SDR plus physical therapy versus physical therapy alone was conducted.[3–6] It has been shown that in a short-term period (up to two years), SDR reduces spasticity, increases a patient's passive range of motion along with positive impacts on gait pattern and gross motor function.[2, 3] Although SDR has been performed for about 30 years, studies reporting the effects of SDR over a long-term period have only appeared recently. Furthermore, possible adverse effects associated with the surgery, such as the development of scoliosis, have been suggested. The aim of the present review was to summarise the outcomes of SDR over a long-term period and to summarise adverse events, such as the development of spinal abnormalities, in children with spastic cerebral palsy that have undergone SDR.

Method

Operational Definitions of SDR and Cerebral Palsy

For the present review, we included studies that described the outcomes and adverse effects in patients undergoing SDR who were younger than 18 years of age at the time of the operation. We included studies in which SDR was performed at the lumbar level. In SDR the dorsal roots are exposed after a multilevel laminotomy/laminectomy or after a single laminectomy in combination with ultrasonographic location of the conus and separated into different rootlets. The rootlets to be transsected are identified with the EMG response, after electrophysiological stimulation, and the palpation of muscle contraction. Details with respect to the operative procedures are provided in table 1. At least 50% of the subjects were diagnosed with cerebral palsy or the outcome results were documented separately for patients diagnosed with cerebral palsy. As we were interested in the long term follow up, we aimed to review the results of studies with a follow-up period of at least five years. For this purpose we included studies with regular follow up examinations performed 5 years or more after SDR or studies which reported a mean follow up duration of at least 5 years. Only articles written in English were included in the review.

Literature Search

The following databases were searched: MEDLINE (Feb 28nd 2010/254 articles), Embase (Feb 28nd 2010/314 articles), Web of Science (Feb 28nd 2010/416 articles), the Cochrane Library of systematic reviews (Feb 28nd 2010/1article) and the Physiotherapy Evidence Database (PEDro; Feb 28nd 2010/5 articles). The search terms used for the target sample were "rhizotomy AND spasticity" as well as "rhizotomy AND cerebral palsy". In total, 759 titles and abstracts were scanned. Six hundred seventy-five articles were excluded because they a) did not report data with respect to SDR or SDR was not performed at the lumbar level, b) the subjects were older than 18 years of age when they received the surgery, c) the studies did not report outcomes and/or adverse events, d) the follow-up period was less than five years and e) the subjects were not diagnosed with cerebral palsy. The full text of the remaining 84 titles was read through in detail. Another 63 articles were excluded because they did not meet the inclusion criteria. In six studies SDR was not performed at the lumbar level, 10 studies included mainly patients older than 18 years of age, in 8 studies SDR was not performed in patients with CP, 10 studies did not report outcomes after SDR and in 63 studies the follow up duration was less than 5 years or the follow up duration was not reported in detail. Twenty-one studies were finally included in the review. Table 1 summarizes the study populations; diagnosis and the diagnostic subgroups; the level of functioning; the age when selective dorsal rhizotomy was performed; details with respect to the operative techniques and the postoperative rehabilitation; the follow-up time of the study; the control group (if present) and the type of intervention that was performed in the control group.

Organisation of the evidence and data extraction

Two authors (SG and RJV) independently read and judged all the identified articles separately according to the criteria mentioned below. Finally, the authors came together, compared their data and reached a consensus.

Level of evidence and conduct of study

When making treatment decisions, it is important to know whether these decisions are based on the results of rigorously controlled investigations, or to know that they based on the results of uncontrolled clinical observations – or on results with even lesser evidence. The level of evidence for each study was coded according to a hierarchy of the levels of research design (see Table 2). Level I designs impose the most experimental control available for an intervention, and if well conducted, produce the most definitive results. Level II and III designs can produce tentative conclusions.

Level IV studies merely suggest causation while no conclusions regarding treatment efficacy can be drawn from Level V evidence. Nevertheless, Levels IV and V studies are important for demonstrating whether or not more robust research is warranted. Studies with designs capable of producing at least tentatively conclusive evidence (Levels I–III) were further organized to analyze the evidence. The quality of the actual conduct of the studies using Level I–III designs were evaluated with the following questions:

1. Were inclusion and exclusion criteria of the study population well described and followed?
2. Was the intervention well described and was there adherence to the intervention assignment? For two-group designs, was the control exposure also well described?
3. Were the measures used clearly described, valid and reliable for measuring the outcomes of interest?
4. Was the outcome assessor unaware of the intervention status of the participants (i.e. were there blind assessments)?
5. Did the authors conduct and report appropriate statistical evaluations including power calculations?
6. Were dropouts/loss in follow-ups reported less than 20%? For two-group designs, were dropouts balanced?
7. Considering the potential within the study design, were appropriate methods for controlling confounding variables and limiting potential biases utilised?

The conduct of the study was rated strong (yes score on 6–7 questions), moderate (score 4–5), or weak (score </= 3). The level of evidence and conduct of study rating is also summarized in Table 1 (Summary of studies).

Type of outcome by International Classification of Functioning, Disability, and Health (Table 3).[7]

Each outcome from the Level I–III studies was classified by whether it had an effect on body parts or function, on functional activities or participation, or in the context of life (i.e., societal or physical barriers, family capacities). These outcomes are summarized in Table 4 and this evidence table is analyzed and discussed in the text. For readers who are interested in outcomes for which there is currently only suggestion of causation or some data that they may warrant further study, Appendix 1 lists the outcomes studied in the Level IV and V studies included in this review.

Adverse events

Finally, the authors summarized the adverse events that have been reported (Table 6).

Discussion

Analysis and discussion of the evidence

In summary the strength and the quality of the evidence with respect to the outcome five or more years after SDR in children with spastic cerebral palsy is very limited. In the present review we included 21 studies, which have been conducted between 1998 and 2010 and included 966 children.[8–28] The study subjects ranged in age from two to 27 years at the time of the SDR. The level of evidence produced by these studies was poor to moderate with 18 Level IV studies[8–11, 13–18, 20–26] unable to evaluate causation and three Level III studies[12, 19, 27] able to provide tentative conclusions that the outcomes were attributable to the SDR.

What evidence exists about the long-term effects of selective dorsal rhizotomy in the body function and body structure components in children with cerebral palsy?

Two level III studies reported outcomes in the domain of body function and body structure at 5 to 20 years after SDR.[12, 27] Subramanian et al. evaluated the gait performance of 11 children with spastic CP 10 years after SDR using 2-dimensional gait analysis. The data were compared with a group of age matched healthy controls. Although an increase of dimensionless step length and gait speed was reported one and three years after SDR, the changes on temperospatial gait parameters could not be maintained 10 years after SDR and were significantly lower than those in healthy controls. However, gait kinematics in knee and hip improved significantly and were maintained 10 years after SDR.[27] Langerak et al. re-evaluated the same study group 10 years later and found that the changes in gait kinematics persisted 20 years after SDR. Furthermore, dimensionless step length and dimensionless cadence increased significantly compared to the preoperative values and the values 10 years after SDR. Twenty years postoperatively the temperospatial parameters in patients undergoing SDR were close to the values of the healthy control group.[12]

What evidence exists about the long-term effects of selective dorsal rhizotomy in the activity component in children with cerebral palsy?

One level III study reported outcomes five years after SDR in the activity domain. In this study the outcomes of 21 children with spastic CP undergoing SDR followed by intensified physical therapy were compared to the outcomes of 21 children diagnosed with spastic CP that were treated with intensified physical therapy alone. Both the intervention group and the control group improvement improved significantly in of function 5 years after SDR. However, the improvement after SDR was not significantly different between children that underwent SDR and children that were treated by intensified physical therapy alone.[19]

What evidence exists about the long-term effects of selective dorsal rhizotomy in the participation component in children with cerebral palsy?

There was no Level III study reporting outcomes in the participation domain 5 years or more after SDR.

What forms and magnitudes of complications have been documented in children with cerebral palsy long after selective dorsal rhizotomy?

Six studies reported adverse events after a follow-up time of more than five years.[8, 11, 13, 15, 26, 28] The follow-up times ranged from 5.8 to 21.4 years. All studies focused on the presence of spinal abnormalities and back pain. In all studies, the incidence of spinal abnormalities was evaluated with frontal and sagittal x-rays of the spine. In one study, an MRI of the spinal cord and the spine was performed in addition to x-rays.[13] Four studies reported the presence of a scoliosis in 41–56% of all children undergoing SDR .[8, 11, 13, 28] Five studies reported a kyphosis in 2–12% of patients that underwent SDR.[8, 11, 13, 15, 28] A lumbar lordosis was described in 10–50% of the patients.[8, 11, 13, 28] A spondylolysis was reported in 7–37% of children after SDR [8, 13, 15] and a grade I spondylolisthesis was reported in 2–24% of the children [8, 11, 13, 28]. Five to 29% of the patients undergoing SDR suffered from back pain at the long-term follow-up.[11, 13, 26, 28] Langerak et al. performed spinal MRIs in a group of thirty patients who received SDR 21.4 years after SDR and reported spinal stenosis in 27% of patients, disc protrusion in 7% and black discs in 20%.[13] No sensory abnormalities or difficulties with bladder/bowel/sexual function were reported.

What is the strength of the evidence?

Strength of a body of evidence depends on the number of studies and the total number of people who have been studies, the strength or quality of the evidence produced, and the consistency of the evidence. In summary, the strength of evidence in the studies included for this review is very poor and the conduct of the studies was weak to moderate. The two level III studies, which supported a positive effect of SDR on gait abnormalities in patients with spastic cerebral palsy ten and twenty years after SDR only included a number of patients and and lacked a power calculation.[12, 27] The outcome assessment was based on two-dimensional gait analysis, which is not the gold standard for the evaluation of gait abnormalities and might not objectively represent gait kinematics.[29] In the third level III study which could not show a causative effect of SDR on gross motor function, no power calculation was reported and it was not stated whether the outcome-assessors of the study were blinded for the type of intervention. Furthermore, this study used outcome measures to assess gross motor function that have not been validated for children with CP or were not developed to assess outcome. Unfortunately the results with respect to spasticity in this study were not provided separately for the children who received physical therapy alone.[19]

Conclusion and Future Directions

SDR is performed routinely in various countries for the treatment of lower-limb spasticity in children with spastic cerebral palsy. Previous studies have repeatedly shown that SDR reduces spasticity and increases passive range of motion, improves gait and leads to an improvement of gross motor function in the short term. The positive effects of SDR on the short term have been summarised in a systematic review and in a meta-analysis.[2, 3] Both studies showed that SDR has a positive influence on the ICF body structure & functions and activity domains.[2, 3] However, data with respect to the long-term effects of SDR were mostly published later. In the present review, we summarised the results of studies with a follow-up time of more than five years that examined the effects of SDR. The review summarises the results of twenty-one studies. Three level III studies[12, 19, 27] and eighteen level IV studies [8–11, 13–18, 20–26, 28] were ultimately included. Level IV studies are not able to attribute any changes observed to the intervention alone. However, because as only few higher level studies could be identified for this review, the results of 11 level IV studies which reported outcomes 5 or more years after selective dorsal rhizotomy in children with spastic CP are summarized in appendix 1 and discussed below.

Two level III studies – including 11 and 13 patients respectively – showed an effect of SDR on the body function and structure domain.[12, 27] The study of Mäenpaa et al – which included 21 patients – was classified as level III study.[19] However, the results in the body function and structure domain were not mentioned separately for the patients in the control group. Six further level IV studies including 244 patients reported outcomes in the domain of body structure and body function.[10, 14, 18, 21, 23, 25] A significant decrease spasticity and range of motion was found was found 5 years after SDR in four studies [10, 18, 19, 21] and in one study 20 years after SDR[14]. One level IV study reported a significant improvement of gait performance in a subgroup of children that have been younger than 5 years old when SDR was performed.[23] There was no association between the age at the operation and outcome in any of the other studies. Whereas the one level III study including 21 patients which assessed outcomes in the activity domain 5 years after SDR could not find a causative relation between SDR and the postoperative improvement of gross motor function[19], eight level IV studies including 240 patients reported outcomes on the domain of activity [9, 10, 14, 17, 18, 20–22] and five of those showed a significant increase of gross motor function 5 to 20 years after SDR [14, 17, 18, 20, 21]. Furthermore one study reported a significant increase of fine motor skills 5 years after SDR.[22] No level III study reported outcomes in the domain of participation, but two level IV studies involving 85 patients assessed outcomes in the participation and reported a significant improvement of self-care indepence five years after SDR[18, 20].

The results of the study of Mäenpää[19], which could not find a causative effect of SDR on gross motor function 5 years after SDR are in contrast to the studies of Subramanian et al and Langerak et al who found a positive effect of SDR on gait performance[12, 27]. Whereas there were no considerable differences in operative technique, postoperative rehabilitation and the age at the operation among these studies, the study subjects differed considerably with respect to their preoperative functional status. Whereas in the study of Mäenpää et al. the mean preoperative Gross Motor Function Classification Scale (GMFCS)[30] level was 3.8 in patients undergoing SDR (indicating that most of the patients were nonambulators or assisted ambulators)[19], the studies of Subramanian et al and Langerak et al included patients who were independent or assisted ambulators (thus GMFCS level scores of I–III)[12, 27]. Therefore question arises whether the preoperative functional status might have an influence on the long term outcome after SDR in children with spastic CP. A correlation between outcome and preoperative functional level was not reported in any of the studies. In the study of Mäenpää et al the intervention group and the control group were not identical with respect to their preoperative functional status.[19] Although the gross motor development differs considerably in individuals with CP,[31] it may be assumed that motor development in children with cerebral palsy stops earlier in children who are more severely involved.[32] The improvements of functional status in the 5 year follow up probably mainly represents spontaneous motor development in the study of Mäenpää et al.[19] However, as the intervention group was more severely affected than the control group and the improvements of gross motor function where better in the intervention group (although not statistical significant, it might still be assumed that SDR could have had a positive impact on gross motor function in the intervention group.

One major limitation in the study of Mäenpää et al is the choice of outcome assessments.[19] When evaluating gross motor function in patients with CP, standardized outcome measures that have been evaluated for this specific patient group should be used. Whereas three of the four Level IV studies, which showed a significant improvement of gross motor skills after SDR used standardized instruments that have been validated for the outcome assessments in children with CP (the Gross Motor Function Measure {GMFM}[33] and the Pediatric Disability Inventory {PEDI}[34])[18, 20, 21], in the study of Mäenpää et al gross motor function was evaluated by the Illinois-St.Louis Scale and the Gross Motor Classification Scale. The Illinois-St.Louis Scale – to our knowledge – has not been evaluated for validity, reproducibility or responsiveness in patients with CP. The GMFCS – which is a is a very valuable tool to classify patients with cerebral palsy with respect to their functioning and has been evaluated for these purposes extensively – was not developed to measure outcomes after an intervention and therefore should not been used as an outcome parameter.[35]

Six studies with a follow-up time of five to twenty years assessed the incidence of spinal abnormalities and/or back pain after SDR.[8, 11, 13, 15, 26, 28] Although a great number of the patients showed evidence of spinal abnormalities in the follow-up, no comparisons were made to a group of patients who did not receive SDR and it remains unclear to what extend SDR is associated with spinal deformities. Spinal deformities – especially scoliosis – are often observed in conjunction with cerebral palsy. Fifteen to eighty percent of patients with CP have a scoliosis. This wide range in prevalence is due to variations in age, nature, and severity of neurological dysfunction, all of which affect the extent of physical impairment in the populations being studied.[36]

In summary, there is poor to moderate evidence that SDR has a positive long-term effect on the ICF body structure and function domains. There is no evidence that SDR has a positive long-term influence on the ICF activity domain and there is no evidence that SDR has a positive influence on the ICF participation domain in children with spastic cerebral palsy. In addition, spinal deformities, such as scoliosis and lordosis, are often observed in patients with cerebral palsy that received SDR. However, it remains unclear to what extent these abnormalities are due to SDR.

Thus, data on the long-term outcome of SDR in children with spastic cerebral palsy are limited and future studies are needed. In general, studies assessing long-term outcomes after an intervention are subject to various methodological difficulties. For example, randomised controlled trials are not suitable for the assessment of long-term outcomes, as the intervention group and the controls might undergo different additional interventions during the follow-up periods. This might influence the outcome and explain the differences in outcome between the groups. An alternative – and maybe more suitable – research design would be a matched controlled study, in which a population that underwent SDR could be compared with a matched population that did not receive the surgery. If longitudinal data of a specific measurement are available, they could be compared with the development of a cohort that receives SDR and the pre-post measurements can be compared with the available growth curves of that specific measurement. With respect to the ICF body structure and function domains, we suggest that future studies include larger patient populations. Gait abnormalities should – whenever possible – be performed by three-dimensional gait analysis. In regards to the ICF activity domain, the conflicting evidence with respect to the gross motor outcome after SDR over a long-term period requires further study. Future studies should assess gross motor function with validated and reproducible methods which are designed to assess outcomes with cerebral palsy. In addition, we suggest that future studies use standardised questionnaires to assess the ICF participation domain. Furthermore, when adverse events after SDR– such as spinal deformities – are reported, these events should be compared to a group of similar

subjects with cerebral palsy who did not undergo SDR. Finally, the question whether preoperative patient characteristics, such as functional status and age at operation might influence the long term outcome after SDR in children with spastic CP should be addressed.

TABLE 1

		Level of Evidence	Conduct Rating	Population	Intervention	Control State	Total N	Ages	
Golan et al.[8]	2007	IV	NA	Spastic CP. 33% NYU I, 53% NYU II, 9% NYU III, 5% NYU IV	Laminotomy (L1-S1). SDR (L2-S2) with electrophysiological guidance. 6 weeks inpatient rehabilitation	NA	98	5.1 y (3–11)	5.8 y (1.1–11.5)
Grunt et al.[9]	2010	IV	NA	Spastic Diplegia. GMFCS I- III	Laminotomy (L2–L5). SDR (L2-S1, max. 50% of rootlets). 12 months intensified physical therapy	NA	18	6.8 y (6–10)	5.5 y (1.1–9)
Gul et al.[10]	1999	IV	NA	Spastic CP	SDR. Electrophysiological guidance. Intensified physical therapy for 9 months	NA	33	4.4 y (2–10)	5 y (4–6)
Johnson et al.[11]	2004	IV	NA	Spastic CP. Diplegia. Ambulatory with/without assistive devices	Laminectomy (L1–L5) or laminoplasty (T12-L5, T12-L4, L1-L5). SDR (L2-S1, max. 48% of rootlets). Electrophysiological and clinical guidance. 4–6 weeks inpatient rehabilitation.	NA	34	6.0 y (4–11)	8.6 y (5–11.6)
Langerak et al.[13]	2008	III	4/7	Spastic CP. 23% Household ambulators, 77% independent ambulators	Laminectomy (L2–S1). Electrophysiological guidance	12 age matched healthy controls	13	7.3 y (2–14)	20 y
Langerak et al.[12]	2009	IV	NA	Spastic CP. 57% GMFCS II, 43% GMFCS III	SDR	NA	14	8 y	20 y
Langerak et al.[14]	2009	IV	NA	Spastic CP; Diplegia. Ambulant with/without assistive devices before age 4 years	Laminectomy (L1/2–S1, L1–L5, L2–L5, L2–S2 and L3–S1). SDR	NA	30	5.2 y (2–27)	21.4 y (17–26)
Li et al.[15]	2008	IV	NA	Not reported	Laminectomy (L2–S1). SDR (L2–S1). Electrophysiological guidance	NA	61	6.9 y (3–20)	6.3 y (5–9)
Lundkvist et al.[17]	2009	IV	NA	Spastic CP; Diplegia. 3% GMFCS I, 23% GMFCS II, 29% GMFCS III, 43% GMFCS IV, 3% GMFCS V	SDR	NA	35	4.5 y (2.5–6.6)	5 y

Lundkvist et al.[16]	2006	IV	NA	Spastic CP; Diplegia. 3% GMFCS I, 22% GMFCS II, 32% GMFCS III, 41% GMFCS IV, 3% GMFCS V	SDR	NA	41	4.4 y (2.6–6.6)	5 y
Mäenpää et al.[19]	2003	III	5/7	Spastic CP; 81% diplegia and 9% quadriplegia. Mean GMFCS 3.8	Laminotomy. SDR (from S1, max. 66% of the rootlets). Electrophysiological guidance. 6 weeks inpatient rehabilitation. Intensive PT for 6 months.	21 age matched patients with CP receiving normal intensity PT	21	6 y (3–11)	5 y
Mittal et al.[20]	2002	IV	NA	Spastic CP; 84% Diplegia, 7% triplegia, 9% quadriplegia. 29% NYU I, 49% NYU II, 20% NYU III, 2% NYU IV. Total study population: n = 45. 5-year follow-up: n=25.	Laminotomy (L1–S2). SDR. Electrophysiological guidance. 6 wks intensified inpatient rehabilitation.	NA	25	4.5 (3.0–7.4)	5 y
Mittal et al.[21]	2002	IV	NA	Spastic CP; 80% Diplegia, 6% triplegia, 14% quadriplegia. 69% NYU I or II, 25.4% NYU III, 5.6% NYU IV. Total study population: n=71. 5-year follow-up: n=50.	Laminotomy (L1–S2). SDR. Electrophysiological guidance. 6 wks intensified inpatient rehabilitation.	NA	50	5.2 y (3–10.7)	5 y
Mittal et al.[22]	2002	IV	NA	Spastic CP; 88% Diplegia, 5% triplegia, 7% quadriplegia. 40% NYU I, 44% NYU II, 15% NYU III, 3% NYU IV. Total study population: n = 41. 5-year follow-up: n=30.	Multilevel Laminotomy. SDR. Electrophysiological guidance. 6 weeks intensified inpatient rehabilitation.	NA	30	4.8 y (3.0–7.5)	5 y
Nordmark et al.[18]	2008	IV	NA	Spastic CP; Diplegia. 3% GMFCS I, 23% GMFCS II, 29% GMFCS III, 43% GMFCS IV, 3% GMFCS V	Laminoplasty (L1-L5). SDR (L2–S2, up to 83% of rootlets).	NA	35	4.5 y (2.5–6.6)	5 y
O'Brien et al.[24]	2005	IV	NA	Spastic CP	Single Laminectomy (T12, L1 or L2). Ultrasonographic location of the conus. Electrophysiological guidance. SDR (L1–S1, 60–70% of rootlets)	NA	158	2–14 y	7.5 y (5–9)

O'Brien et al.[23]	2004	IV	NA	Spastic CP. 65% not walking, 35% walking with walker	Single Laminectomy (L1, L2). Ultrasonographic location of the conus. Electrophysiological guidance. SDR (L1–S1, 60–70% of rootlets)	NA	52	2–15 y	7.5 y (5–9)
Salame et al.[25]	2003	IV	NA	CP, multiple sclerosis, trauma, degenerative disease. Subgroup of 60 patients with spastic CP	Single laminectomy L1 (L2). SDR (L1-S1). Electrophysiological guidance only in some cases.	NA	60	12 y	11 y
Spiegel et al.[27]	2004	IV	NA	Spastic CP; 68% diplegic, 15% triplegic, 16% quadriplegic. 81% community ambulators, 10% household ambulators, 8% therapeutic ambulators, 1% nonambulators.	Laminoplasty. SDR (L1–S2, 16%–61% of rootlets).	NA	79	6.2 y (SD 1.8)	5.8 y (SD 2.4)
Subramanian et al.[27]	1998	III	3/7	Spastic CP. Ambulant	Laminectomy (L2–S1). SDR (L2–S1). Electrophysiological Guidance	12 age matched healthy controls	11	7.8 y (2.5–13.2)	10.6 y (10.4–11)
Turi et al.[28]	2000	IV	NA	Spastic CP. 21% community ambulators (n=10); 6% household ambulators , 9% therapy ambulators, 64% nonambulators	Laminectomy (L2–S1 and S2). SDR (L2–S2, 30–100% of rootlets)	NA	47	6.8 (2.25–30)	5.3 y (2–9)

Abbreviations used: CP=Cerebral Palsy, NYU = New York University Scale, SDR = Selective Dorsal Rhizotomy, GMFCS = Gross Motor Function Classification

TABLE 2 **Hierarchy of the levels of research design**

Level of Evidence	Type of Study
I	Systematic review of randomised controlled trials (RCTs) Large RCT (with narrow confidence intervals) (n > 100)
II	Smaller RCT's (with wider confidence intervals) (n<100) Systematic reviews of cohort studies "Outcomes research" (very large ecologic studies)
III	Cohort studies (must have concurrent control group) Systematic reviews of case control studies
IV	Case series Cohort study without concurrent control group (e.g. with historical control group) Case-control Study
V	Expert Opinion Case study or report Bench research Expert opinion based on theory or physiologic

TABLE 3 **Components of the World Health Organization's International Classification of Functioning, Disability and Health (ICF)**

ICF Domain	Definition
Body Structures and Body Functions	*Body Structures* are anatomical parts of the body such as organs, limbs and their components. Impairments are problems in body structure as a result of deviation or loss. *Body Functions* are the physiologic functions of body systems. Impairments are problems in body function as a result of deviation or loss.
Activity and Participation	*Activity* is the execution of a task or action by an individual. Activity limitations are difficulties an individual may have in executing activities. *Participation* is involvement in a life situation. Participation restrictions are problems an individual may experience in involvement in life situations.
Contextual Factors	*Environmental factors* make up the physical, social and attitudinal environment in which people live and conduct their lives. They can be viewed as facilitators (positive influence) or barriers (negative influence). *Personal factors* are the particular background of an individual's life and living and compromise features of the individual that are not part of a health condition.

TABLE 4 **Study outcomes according to the domains of the International Classification of Functioning, Disability and Health**

Study	Outcome of interest	Measure	Domain	Result
Langerak et al.[12]	Knee ROM	2d Gait Analysis	BS/BF	SS (p<0.0001)
	Hip ROM	2d Gait Analysis	BS/BF	SS (p=0.04)
	Knee Midrange value	2d Gait Analysis	BS/BF	NS
	Hip Midrange Value	2d Gait Analysis	BS/BF	NS
	Dimensionless Cadence	2d Gait Analysis	BS/BF	SS (p=0.003)
	Dimensionless Step Length	2d Gait Analysis	BS/BF	SS (p=0.02)
	Knee ROM	2d Gait Analysis	BS/BF	SS (p<0.001)
	Hip ROM	2d Gait Analysis	BS/BF	NS
	Knee Midrange value	2d Gait Analysis	BS/BF	SS (p<0.001)
	Hip Midrange Value	2d Gait Analysis	BS/BF	SS (p<0.001)
	Dimensionless Cadence	2d Gait Analysis	BS/BF	SS (p=0.005)
	Dimensionless Step Length	2d Gait Analysis	BS/BF	SS (p=0.002)
Mäenpää et al.[19]	Spasticity Hip Flexors	MAS	BS/BF	SS (p<0.05)
	Spasticity Hip Rotators	MAS	BS/BF	SS (p<0.05)
	Spasticity Hip Adductors	MAS	BS/BF	SS (p<0.05)
	Spasticity Knee Flexors	MAS	BS/BF	SS (p<0.05)
	Spasticity Plantar Flexors	MAS	BS/BF	SS (p<0.05)
	Gross Motor Function	Illinois-St.Louis Scale	A	SS (p<0.05)
	Gross Motor Function	GMFCS	A	NS
	Knee ROM	2d Gait Analysis	BS/BF	NS
Subramanian et al.[27]	Hip ROM	2d Gait Analysis	BS/BF	NS
	Knee Midrange value	2d Gait Analysis	BS/BF	SS (p<0.05)
	Hip Midrange Value	2d Gait Analysis	BS/BF	SS (p<0.05)
	Dimensionless Cadence	2d Gait Analysis	BS/BF	SS (p<0.05)
	Dimensionless Step Length	2d Gait Analysis	BS/BF	SS (p<0.05)
	Gait Velocity	2d Gait Analysis	BS/BF	SS (p<0.05)
	Knee ROM	2d Gait Analysis	BS/BF	SS (p<0.05)
	Hip ROM	2d Gait Analysis	BS/BF	NS
	Knee Midrange value	2d Gait Analysis	BS/BF	SS (p<0.05)
	Hip Midrange Value	2d Gait Analysis	BS/BF	SS (p<0.05)
	Dimensionless Cadence	2d Gait Analysis	BS/BF	SS (p<0.05)
	Dimensionless Step Length	2d Gait Analysis	BS/BF	SS (p<0.05)
	Gait Velocity	2d Gait Analysis	BS/BF	SS (p<0.05)

Abbreviations used in TABLE 4: ROM = Range of Motion, GMFCS = Gross Motor Function Classification Scale, MAS = Modified Ashworth Scale, BS = Body Structure and Function, A = Activity, SS = Statistically significant, NS = Not significant.

TABLE 5 **Reported adverse events after Selective Dorsal Rhizotomy**

Study	Scoliosis	Kyphosis	Lordosis	Spondylosis & Spondylolisthesis	Other
Golan et al.[23]	After SDR: 45%	After SDR : 12%	After SDR : 33% Pre-Post-Analysis: NS	Spondylosis after SDR: 12% Spondylolisthesis after SDR: 6%	NR
Johnson et al.[24]	Before SDR: 21% After SDR: 41% Progression > 5°: 6% Pre-Post Analysis: NS	Before SDR: 0% After SDR: 9% Pre-Post Analysis: NS	<20° before SDR: 50% > 50°after SDR : 50% Pre-Post Analysis: p=0.0001	Spondylolisthesis before SDR: 6% Spondylolisthesis after SDR: 24% Pre-post Analysis: NS	Back Pain: 29%
Langerak et al.[25]	Before SDR: 0 After SDR: 56% Pre-Post Analysis: p < 0.01	Before SDR: 0 After SDR: 7% Pre-Post Analysis: NS	Before SDR: 20% After SDR: 40% Pre-Post Analysis: NS	Spondylolysis after SDR: 37% Spondylolisthesis after SDR: 3% Pre-Post analysis: NS	Back Pain: 23% Spinal Stenosis : 27% Disc Protrusion: 7% Black Disc: 20%
Li et al.[26]	NR	After SDR: 2%	NR	Spondylolysis after SDR: 7% Spondylolisthesis after SDR: 7%	NR
Spiegel et al.[27]	Follow Up < 5 y	Follow Up < 5 y	Follow Up < 5 y	Follow Up < 5 y	Back Pain: 5%
Turi et al.[28]	Before SDR: 7% After SDR: 45%	After SDR: 5%	After SDR: 10%	Spondylolisthesis: 2%	Back Pain: 9% Spinal Surgery: 6%

APPENDIX 1 **Other outcomes or Outcomes from Less Robust Studies or Outcomes from Level IV Studies**

Study	Outcome of interest	Measure	Domain	Result
Grunt et al.[9]	Gross Motor Function	GMFM-66	A	NR
Gul et al.[10]	Hip adductors spasticity	MAS	BS/BF	SS (p<0.005)
	Quadriceps Strength	MRC	BS/BF	SS (p=0.002)
	Hip Abduction ROM	Goniometry	BS/BF	SS (p=0.0004)
	Hamstrings Spasticity	MAS	BS/BF	NR
	Ankle Plantarflector Spasticity	MAS	BS/BF	NR
	Knee Extension ROM	Goniometry	BS/BF	NR
	Ankle Dorsiflexion ROM	Goniometry	BS/BF	NR
	Hip Abductor Strength	MRC	BS/BF	NR
	Ankle Dorsiflector Strength	MRC	BS/BF	NR
	Hip Extensor Strength	MRC	BS/BF	NR
	Motor Function	Motor Function Score	A	NR
Langerak et al.[14]	Muscle Tone	5 Point ordinal Scale	BS/BF	SS (p<0.001)
	Joint Stiffness	4 Point ordinal Scale	BS/BF	SS (p=0.019)
	Voluntary Movement	5 Point Ordinal Scale	BS/BF	SS (p=0.002)
	Rolling	4 Point ordinal Scale	A	SS (p=0.004)
	Side Sitting	5 Point Ordinal Scale	A	NS
	Long Sitting	5 Point Ordinal Scale	A	SS (p=0.001)
	Kneel Standing	5 Point Ordinal Scale	A	SS (p=0.002)
	Prone Kneeling	5 Point Ordinal Scale	A	SS (p=0.005)
	Half Kneeling	5 Point Ordinal Scale	A	SS (p=0.003)
	Crawling	5 Point Ordinal Scale	A	SS (p=0.008)
	Standing	5 Point Ordinal Scale	A	SS (p=0.028)
	Walking	5 Point Ordinal Scale	A	SS (p=0.05)
Lundkvist et al.[17]	Gross Motor Function	GMFM88	A	NR
	Gross Motor Function	GMFM88 Goal Score	A	NR
	Gross Motor Function	GMFM 66	A	NR
Mittal et al.[22]	Fine Motor Skills	Fine Motor Scales of PDMS	A	SS (p<0.001)
	Fine Motor Skills	PDMS Total Raw Score	A	SS (p<0.001)
	Fine Motor Skills	PDMS Age Equivalent	A	SS (p<0.001)
	Fine Motor Skills	PDMS Percentile Score	A	NR
	Fine Motor Skills	PDMS z Score	A	NR
	Gross Motor Function	GMFM-66	A	SS (p<0.001)
Mittal et al.[21]	Hip Adductor Spasticity	NYU Tone Scale	BS/BF	SS (p<0.001)
	Hip Abduction ROM	NYU Tone Scale	BS/BF	SS (p<0.001)
	Standing	GMFM 88 Dimension D	A	SS (p=0.001)
	Walking, Running, Jumping	GMFM 88 Dimension E	A	SS (p=0.001)
	Alignment	NYU Rhizotomy Evaluation Form	BS/BF	SS (p<0.001)
	Self Care	PEDI Functional Skills Self Care	A & P	SS (p<0.001)
Mittal et al.[20]	Mobility	PEDI Functional Skills Mobility	A & P	SS (p<0.001))

Nordmark et al.[18]	Gross Motor Function	GMFM88	A	SS ($p<0.001$)
	Gross Motor Function	GMFM88 Goal Score	A	SS ($p<0.001$)
	Muscle Tone Hamstrings	MAS)	BS/BF	NR
	Muscle Tone Hip Adductors	MAS	BS/BF	NR
	Muscle Tone Ankle plantar Flexors	MAS	BS/BF	NR
	Tendon Reflexes lower extremities	NINDS scale	BS/BF	SS ($p<0.001$)
	Tendon Reflexes upper extremities	NINDS scale	BS/BF	SS ($P<0.01$)
	PROM hip adduction	Goniometry	BS/BF	SS ($p<0.001$)
	PROM popliteal angle	Goniometry	BS/BF	SS ($p<0.001$)
	PROM ankle dorsiflexion	Goniometry	BS/BF	SS ($p<0.001$)
	Self Care	PEDI Functional Skills Self Care	A & P	SS ($p<0.001$)
	Mobility	PEDI Functional Skills Mobility	A	SS ($p<0.001$)
	Self Care	PEDI Caregiver Assistance Self Care	A & P	SS ($p<0.001$)
	Mobility	PEDI Caregiver Assistance Mobility	A	SS ($p<0.001$)
O Brien et al.[23]	Gait[a]	12 Point Ordinal Scale	BS/BF	SS ($P<0.05$)
	Gait[b]	12 Point Ordinal Scale	BS/BF	NS
Salame et al.[25]	Spasticity	MAS	BS/BF	NR

[a]Patients younger than 5 years when SDR was performed. [b]Patients older than 5 years when SDR was performed. ROM, Range of Motion; GMFM, Gross Motor Function Measure; MRC, Medical Research Council Scale; NYU , New York University Scale; GMFCS, Gross Motor Function Classification Scale; MAS, Modified Ashworth Scale; PDMS , Peabody Developmental Measure Scale; PEDI, Paediatric Disability Inventory; NINDS, National Institute of Neurological Disorders and Stroke; BS/BF, Body Structure and Function; A, Activity; P, Participation; SS, Statistically significant; NS, Not significant; NR, not reported

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Chapter 8

General Discussion

Introduction

The primary focus of this thesis was to investigate outcomes after selective dorsal rhizotomy (SDR). Amongst others, we aimed to describe the outcome after SDR on the level of impairment and activity in children who suffered from bilateral spasticity due to other etiologies than spastic CP. Illustrated by a clinical report of two cases we discussed whether SDR might represent a treatment option for the management of lower limb spasticity in patients who suffer from neurodegenerative disease. Furthermore we aimed to assess whether neuroimaging would be helpful in the selection process for SDR and investigated whether there is a relationship between preoperative MRI findings with the change in functioning after SDR. In addition we aimed to describe the improvement of gait abnormalities after SDR. We addressed the question whether there is a relationship between the preoperative level of functioning and the improvement of gait performance. For the assessment of gait abnormalities we used a method to measure sagittal joint and segment angles on video screen. As the clinical utility of this method has not been investigated before, we aimed to assess its reliability and validity. At last we described the effects and adverse events of SDR on long term in a systematic review. The present chapter generally discusses the results of this thesis in the context of its original aims and in the context of previous publications. We also discuss methodological considerations and emphasize the limitations of the thesis. Furthermore, we also consider its clinical implications and provide recommendations for future research.

Methodological considerations

Study population

The present thesis includes two prospective cohort studies. The study population included all patients who underwent SDR at the lumbar level for the treatment of lower limb spasticity in the VU Medical Center, Amsterdam, the Netherlands between January 1998 and March 2009. No control group has been assessed. Only patients with bilateral spasticity were operated. In patients who underwent SDR for the improvement of gait performance, the selection for SDR was based according to definite eligibility criteria. In total 44 patients with bilateral spasticity and walking ability underwent SDR. The study design and lost to the follow up assessments unfortunately did not allow to include all of these 44 patients to be included in both of the prospective cohort studies. Though, the sample sizes were quite small. Whereas with respect to the eligibility criteria the study participants in the two prospective studies were quite homogeneous, there was a heterogeneity with respect to age (ranging from 2.75 years to 13.16 years) and gross motor function level (including patients with gross motor function levels I to III) as well as in the underlying cause of the spasticity.

Furthermore the thesis comprises one methodological study which assessed 17 children with spastic CP. The in- and exclusion criteria in the methodological study differed from those in the prospective studies assessing outcomes after SDR. There were differences with respect to age, gross motor function level and the distribution of the spasticity. In the methodological study only children who walked without assistive devices were examined and the participants suffered from bilateral and unilateral spastic CP. At last a case report which describes the outcome after SDR in two patients diagnosed with progressive neurological disease is part of this thesis.

Treatment

The same neurosurgeon performed the SDR in all children. A laminotomy of L2 to L5 was performed and the dura was opened. Then the dorsal roots L2 to S1 were exposed. Each dorsal root was separated into 3 to 4 different rootlets. Then each rootlet was stimulated by electrostimulation. The transsection of the rootlets was performed according to EMG response and according to the reaction in manual muscle palpation. No more than 50% of the rootlets were transsected at each level. Rootlets/fascicles showing electrical responses upon stimulation of the penis/clitoris were spared to prevent sexual and bladder disturbances. The post-operative rehabilitation included intensive physical therapy (3 to 5 hours per week) for 12 months, which was mostly performed within an outpatient setting.

Preoperative assessment

In all patients who underwent SDR an extensive multidisciplinary assessment was performed preoperatively. The assessment included a thorough orthopedic examination performed by a pediatric physiatrist, a neurological examination performed by a pediatric neurologist, a standardized assessment by a physical therapist and gait analysis. Spasticity was tested with the spasticity test [SPAT] [1], which represents a modification of the Tardieu Scale. Gross Motor Function was classified according to the GMFCS [2]. Furthermore the preoperative evaluation included radiologic examination of the spine (anterior-posterior plane to exclude scoliosis and lateral plane to exclude lordosis, kyphosis as well as spondylosis/spondylolisthesis) as well as an X-ray of the pelvis (anterior posterior plane to exclude subluxation of the hips).

Neuroimaging

Neuroimaging of the brain (and in some cases the spinal cord) was recommended before SDR in all patients. In most of the cases the brain MRI was performed at the VU Medical Center according a white matter protocol, however in some of the patients MRI had already been performed in other centers and therefore was not repeated. The MRIs were classified into three diagnostic groups: periventricular leucomalacia (PVL), hydrocephalus and normal MRI. In the patients with PVL the severity of the MRI abnormalities was graded according to a scoring system

that has been used previously [3]. The following items were assessed: ventricular size, evidence and extension of white matter signal intensity, evidence and extension of white matter loss, thinning of the corpus callosum, dimension (size) of sub-arachnoidal space, evidence and size of cysts, and presence of grey matter abnormalities. The items were scored on a 3-point scale, with a score of 3 indicating the most severe MRI abnormalities. The scores were summed to obtain a total score for each child (minimal score = 7, maximal score = 21). Kappa Scores for the intrarater agreement of the grading scale ranged from 0.87 (white matter signal intensities) to 1.0 (white matter loss, cysts, thinning of the corpus callosum, subarachnoidal space and grey matter abnormalities). Kappa Scores for the interrater agreement ranged from 0.45 (white matter loss) to 1.0 (subarachnoidal space and the grey matter abnormalities).

Outcome Assessment

All patients undergoingwho went SDR at the VU medical centre have been followed yearly including spasticity assessment, orthopedic examination, gait-analysis and gross motor function assessment. Gross Motor function was measured with the GMFM. The GMFM is an observational measure, which has been specifically designed to assess gross motor function in children with CP. Its validity, reliability and responsiveness have been evaluated in children with spastic CP [4]. In this thesis we used the GMFM-66 version. The GMFM-66 provides a numerical scale ranging from 0 to 100 and covers five gross motor dimensions (lying and rolling/crawling and kneeling/standing and walking/running/jumping). The GMFM-66 can be calculated with the Gross Motor Ability Estimator and addresses the linearity of the individual item scores across the entire range of the GMFM scores [5].

Gait analysis was performed in a gait laboratory. The subjects walked barefoot along a 10-m walkway at a self-selected, comfortable speed. Video recordings of both legs were taken in the sagittal and frontal plane. A custom-made, open-source software package that allows observing gait parameters and video recordings (the MoXie Viewer®, www.smalll.nl) was used for the data analysis. This software allows to freely selecting the video speed (including slow motion and freeze-frames) and includes a graphical multi-goniometer to measure six sagittal angles. The sagittal knee angle kinematics were measured before and after SDR by the same observer. We have shown that the measurement of sagittal knee angles shows good intraobserver reproducibility and allows to detect relevant changes in knee angle kinematics [6]. To measure the overall gait performance before and after SDR the Edinburgh Gait Assessment Scale (EGAS) was used. The EGAS is an observational gait analysis scale, which describes gait deviations with 17 different items on an ordinal scale (assessing movements in the foot/ankle, knee, hip, pelvis and trunk) [7].

Previous studies have reported good intra- and interobserver reliability for the EGAS in patients diagnosed with spastic CP [7;8]. Furthermore surface EMG was registered in different muscle groups. EMG profiles were assessed and classified in a qualitative way by two observers, using explicit criteria based on patterns associated with spasticity or co-contraction. In addition, the amount of abnormal activity in the EMG signals were also assessed in a quantitative way.

SDR as a treatment option in children who suffer from spasticity due to other etiologies than CP

SDR has mainly been described as a treatment option to reduce spasticity in children diagnosed with bilateral spastic CP. In an editorial Turner wrote, that – in his experience – SDR has not been shown to be effective in treating hypertonicity of spinal origin, such as spinal cord injury (SCI) and hereditary spastic paraparesis (HSP) and that a good initial decrease in tone occurs, but the hypertonia returns within several years. Furthermore he mentioned, that SDR has also not proved effective in treating the hypertonicity arising from traumatic brain injury (TBI) and anoxia [9]. In contrast various articles – mainly case reports and small case series – reported the effectiveness of SDR in patients who suffer from bilateral spasticity due to other etiologies than CP. Mainly patients who suffered from non-progressive diseases were reported and these reports mainly included adult patients and very heterogeneous diagnostic subgroups.

Good results after SDR were reported in patients with spasticity due to spinal cord injuries or diseases of the spinal cord – such as traumatic SCI [10–14], transverse myelitis [12;13;15] and myelomeningocele [13]. In our report we described good improvements of gross motor function after SDR in a patient with encephalomyelopathy associated with congenitally acquired HIV Infection and another patient who suffered from bilateral spasticity due to a spinal process [16]. In accordance with previous studies [10–15], we found a disappearance of the spasticity in both cases and the spasticity did not reoccur. Both patients dramatically improved their gross motor skills [16]. Although there might be indications that SDR positively influences the outcome in patients with spasticity from spinal origin – our reports and those of others do not allow a causative conclusion.

Furthermore various reports have been published which described the outcomes after SDR in patients who suffered from spasticity originating from a non-progressive lesion in the brain, which occurred after the neonatal period – such as ischemic cerebrovascular injury [11;17], TBI [14;18], cerebral hemorrhage [12], hydrocephalus [13], cerebral hypoxia due to near drowning or heart surgery [13;19], arterio-venous malformation[18] or encephalitis [19]. In summary the short term results have been reported to be favorable. However, compared to patients with spastic CP it appeared that the reduction of the spasticity was less remarkable [19] and that on long term less good effects were described [14]. In our series we have included two patients who suffered from hydrocephalus. One patient was a former preterm child who suffered from post-hemorrhagic hydrocephalus, the other patient showed aqueductal stenosis which was only diagnosed in the preoperative evaluation before SDR. Both patients did not show an improvement of their gross motor abilities after SDR [16]. No other patients with brain lesions that occurred after the neonatal period were treated with SDR in our institution. Despite the positive reports with respect to outcome after SDR in patients who suffer from spasticity due to brain lesions which occurred after the neonatal period, the effect of SDR remains non-conclusive. Furthermore we do not recommend SDR in patients who suffer from hydrocephalus.

SDR has also been performed in patients with progressive neurological disease, such as multiple sclerosis [12], spinocerebellar disease [14] and amyotrophic lateral sclerosis [14]. Furthermore Morota et al reported the outcomes after SDR in patients with degenerative diseases, but did not specify the diagnosis [19]. Whereas an initial improvement was observed in patients with progressive neurological disease, a partial loss of the initial improvement after SDR was observed in the long-term follow up [14]. Furthermore it was mentioned that – compared to patients with spastic CP – the reduction of the spasticity was less remarkable in patients with degenerative disease [19]. We have found a decrease of spasticity after SDR in children who suffer from neurodegenerative disease – leading to improved caregiving and improvements in posturing [20].

Cole et al mentioned one patient with HSP that was treated by SDR but did not show the results separately for this patient [21]. We have not been able to publish the results of SDR in patients with approved HSP. However, we described a decrease of spasticity and a good improvement of functional skills in 4 patients with normal MRI of the brain [16]. Spasticity occurred early in the development in all cases and the lower limbs were mainly involved. Metabolic investigations did not reveal any abnormalities and MRI of the spinal cord was normal as well. Although in none of these cases the family history was positive, HSP was suggested as the most plausible cause of the spasticity. In HSP the genetic evaluation involves mutational screening for various genes, is often protracted and can remain inconclusive. Even though diagnostic

algorithms have been published for the genetic investigation of children with suspected HSP [22], in clinical practice these are often laborious and the results remain inconclusive. Genetic investigations were only partly performed and in none of the patients mutational screening revealed a definite diagnosis. We have treated another patient with normal MRI of the brain and suspected HSP by SDR. She was excluded from the data analysis as a preoperative GMFM assessment could not be performed due to poor compliance. She had a positive family history; her mother and her older sister both suffer from progressive spasticity probably due to an autosomal dominant form of HSP. Unfortunately genetic confirmation of the diagnosis was not available. This patient underwent SDR at the age of 2 years and after SDR the spasticity was permanently reduced and did not reoccur to date (10 years after SDR). She dramatically improved her gross motor function and gait performance.

Does neuroimaging help to select patients for SDR?

The selection criteria to undergo SDR vary considerably from place to place. The first detailed descriptions of selection criteria for SDR are based on the initial studies performed in South Africa [23–25]. Those guidelines included various patient characteristics such as age, diagnosis, tone, ambulatory ability, birth history, motor control, specific medical conditions, orthopedic status, availability of postoperative therapy, and intellectual development [23–26]. The selection criteria to perform SDR in the VU University Medical Center have been mentioned previously [16;27]. In summary many studies reporting outcomes after SDR did not describe selection criteria in detail. In those studies which described the selection criteria, there was a considerable variability between different centers or even between different studies performed by the same study group.

Cerebral palsy is defined as a group of permanent disorders of the development of movement and posture, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain [28]. Neuroimaging studies can help to understand the pathophysiological process which leads to a specific motor disturbance and differing patterns of cerebral lesions or malformations, which are related to the timing of the injury during the development of the brain can be detected with the aid of MRI in children with CP [29–31]. MRI of the brain plays an important role in the diagnostic evaluation of children suffering from spasticity and possibly treatable causes (such as hydrocephalus or vascular malformations) as well as progressive causes of spasticity might be detected with the help of MRI.

Therefore the Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurology Society both advise Magnetic Resonance Imaging (MRI) of the brain for all CP patients [32]. In patients with bilateral spastic CP, MRI of the brain often shows periventricular white matter lesions, mainly observed in former preterm infants [30;31]. In the preoperative evaluation for SDR only few studies included MRI of the brain as a selection criterion [19;33–36]. Some studies only included patients with clear evidence of PVL on neuroimaging for SDR [33;34]. Furthermore evidence of damage to the basal ganglia represented a contraindication in 2 studies [35;37]. None of these studies have performed a subgroup analysis and analysed the outcomes after SDR with differing patterns of cerebral injury.

In chapter 4 we therefore have assessed and classified all available MRIs of the brain in patients with bilateral spastic paresis that underwent SDR to improve walking ability in our centre [16]. Whereas in a previous report of our centre only patients with evidence of PVL on neuroimaging were included for SDR [34], PVL on neuroimaging was not a selection criterion in our further analysis [16]. We have observed differences in outcome between patients with differing types of cerebral lesions. Whereas patients with normal MRI findings showed the best functional improvements after SDR, children with hydrocephalus did not show a functional improvement and children with PVL showed intermediate results. In patients with PVL the severity of the PVL, as well as the single items of a scoring system (ventricular size, white matter signal intensity, white matter loss, thinning of the corpus callosum, cysts, gray matter abnormalities and enlargement of the subarachnoidale space) did not correlate with functional outcome after SDR [16].

The selection process for SDR is complex, time consuming and depends on many other criteria than the results of neuroimaging studies. The decision whether SDR should be performed or not depends on different criteria, such as the age of the patient, type and distribution of the motor disturbance, severity of the disability, gait performance, cognitive abilities, cooperation of the patient and comorbidities. Furthermore a multidimensional and multiproffessional assessment – which follows clearly defined guidelines and includes a thorough orthopedic and neurological examination, an assessment by physical therapists and gait analysis – is mandatory in all patients who are candidates for SDR. Table 1 provides a comprehensive overview of single criteria that have been mentioned in the selection process for SDR in children among different studies. It is beyond of the topic of this thesis to comment on all the criteria that have been defined by different centers in detail.

Is there is a relationship between the preoperative level of functioning and the improvement of gross motor function or gait performance after SDR?

In rehabilitation medicine interventions should be focused on treatment goals. However, it was shown that the definition of treatment goals on short term is not necessarily easy to perform and that the integration of the needs and principal problems of children with their rehabilitation goals may not be optimal [38]. Whereas, already on short term, goal setting represents a considerable challenge for clinicians, focusing treatment goals on long term expectations is – if at all feasible – even more difficult. Nevertheless, the expectation of an intervention should be based on realistic expectations and be in accordance with the needs of the patients and/or his/her surrounding.

With respect to SDR defining specific treatment goals is of particular interest, as it represents an irreversible procedure. The selection of the ideal candidate for SDR is crucial. Only those patients who profit most from SDR should be operated. This process is complex and demanding and the selection of patients depends largely on the expectations of patients, parents, therapists and clinicians. It was repeatedly stated that the ideal candidate for selective dorsal rhizotomy would be a mobile and motivated child who is attempting to improve his/her gait pattern [23;25]. However, many study groups performed SDR also in non-ambulatory patients (though GMFCS levels of IV and V) [19;23;39–42]. Whereas in ambulatory patients with GMFCS levels I-III the main treatment goal is the improvement of motor function, in the non-ambulatory patients with GMFCS levels IV or V the intention of SDR is the improvement of comfort and patient care [25]. When McLaughlin et al. published their meta-analysis in 2002, they stated that it should be speculated that SDR might be most effective for a child whose functional level falls into GMFCS levels III and IV as children with more severe CP may have more potential gain from an invasive procedure. Yet, no statistical analysis was performed to confirm this consideration [43]. In the RCT of Wright et al., which – of the three RCT's included in the meta-analysis – showed the most significant improvement after SDR, the most severely disabled patients were included. In a subgroup analysis however, no significant correlations between the baseline GMFM, age and tone and the GMFM 12 months after SDR were found [44].

In contrast to the statement of McLaughlin et al. [43], Mittal 2005 et al found that 3 and 5 years after SDR patients with milder motor deficits (independent and dependent ambulators) were more likely to improve in functional performance than those who were unable to walk preoperatively [45]. Kim et al performed a large retrospective study and assessed predictors of poor outcome after SDR in an univariate and a multivariate analysis. Poor outcome was defined according to an interview with caregivers (no improvement in lower limb tone, overall motor function, or activities of daily living or a deterioration in overall motor function or ADL were considered as poor outcome). They found that the type of CP (diplegia vs tetraplegia), intellectual delay and mobility (wheelchair vs. crawling vs walking) were significant predictors of outcome in the univariate regression analysis, but only the type of CP retained significant predictive power in the multivariate analysis. Although a poor outcome was observed less frequently in children with GMCS levels I, II and III compared to those in GMFCS level IV and V, this difference was not statistically significant [42].

When the improvement of motor function was the intention of SDR, we only selected patients in GMFCS levels I, II or III for SDR and spasticity needed to significantly interfere with walking performance [16;27]. The interference of spasticity with gait performance was based on clinical examination and the use of gait analysis (including observational gait analysis, kinematic measurements on video screen and surface EMG). We have shown that – with respect to gait performance – the improvement after SDR is significantly better in children who are less severely involved [27]. In contrast, in a large retrospective study Trost et al. could not find a similar difference. The only outcome parameter that differed between groups was gait speed, which was more improved in the patients in GMFCS level III. However, as patients in GMFCS level II already walked significantly faster before SDR possible gains in speed in the higher functioning children might be limited [33]. As mentioned before, there is no international consensus on the preoperative assessment and the selection of patients for SDR. As the selection criteria for SDR considerably differ from place to place, the similarity of study populations with respect to muscle function (e.g. spasticity, strength and selective motor control) between single studies might differ and therefore limit the comparison of different studies. In contrast, it is very likely that there are differences in selection of children for SDR. In walking children, the underlying voluntary muscle strength, the degree of selective motor control and the level of spasticity (defined as a velocity-dependent increase of the tonic stretch reflex) might considerably influence the outcome after SDR. In the Trost. et al study – as in many other studies dealing with SDR – spasticity was assessed by the modified Asworth Scale (MAS). However, the MAS does not correspond with the concept of spasticity – defined as a velocity dependent increase of the tonic stretch-response [46] – and it was stated that its validity and reliability are insufficient to be used as a cinical measure of spasticity [47]. We selected candidates for SDR when spasticity was

present bilaterally and predominantly in the lower limbs. Patients qualified for SDR if the spasticity was present in at least six muscle groups of the lower limbs and was interfering with walking ability. For the assessment of spasticity we used the Spasticity Test (SPAT)[48;49], which is a modification of the Tardieu Test and takes into account that spasticity is velocity dependent and has recently been compared to biomechanical spasticity measurements [48]. In addition the assessment of strength and selective motor control differed between the study of Trost et al.[33] and ours [27] though making a comparison between the two studies is difficult.

Long term outcome after SDR

Although various studies have shown that SDR reduces spasticity and improves gross motor function on short term, only few studies assessed the long term results in patients that underwent SDR. The majority of these studies have been summarized in a systematic review which is part of this thesis [50]. We have found that there is moderate evidence that SDR has a positive long-term influence on the ICF body structure and body function domains but there is no evidence that SDR has an influence on the ICF activity and participation domains.

Since the publication of our review – to our knowledge – three further studies assessed the long term effects of SDR. Langerak et al. assessed the level of activity and participation 17 to 26 years after SDR. They used a semi-structured interview as well as the "The Functional Mobility Scale" and "Life-Habit" questionnaire to gather data on patients' characteristics and long-term experiences after the operation. They concluded that at long-term adults with spastic diplegia undergoing SDR showed high levels of functioning, and similar levels of satisfaction with life habits. In addition the majority of the patients had positive feelings about the neurosurgical procedure. Langerak et al suggested that there is a need for better follow-up after subjects leave school. [51] Tedroff et al assessed the outcome with respect to spasticity, gross motor function, gait performance and passive range of motion 10 years after SDR. There was a reduction in spasticity, however, in some cases the spasticity reoccurred in the knee and ankle joint. With respect to gross motor function they found that the initial improvement after the operation could not be maintained 10 years after SDR. In addition the ambulatory status was best 3 years after SDR and then declined again. A large percentage of the patients undergoing SDR needed orthopedic surgery after the intervention [52]. In contrast to the findings of Tedroff et al. our own study group found a maintenance of the improvements of gross motor function 6 years after SDR but a large percentage of the children still needed additional surgery or botulinum toxin A treatment after SDR [53]. In contrast to previous studies our results were though more positive in GMFM in comparison. However, as mentioned

above, a comparison between studies seems particularly difficult as the selection criteria for SDR vary between studies. Though it remains unclear to what extent differences in the preoperative patient characteristics might help to explain the differences in outcome.

With respect to the adverse events on long term after SDR – it has been repeatedly shown that spinal deformities seem to be common in patients undergoing SDR.[50;54–58] In contrast our personal experience show spinal deformities less commonly in the long-term follow up after SDR. However, the frequency of spinal deformities after SDR has not been systematically analyzed in our own cohort. Question arises whether differences in preoperative patients characteristic – such as the presence of truncal hypotonia and the severity of gross motor disturbance – or the operative techniques might explain the differences between different populations. Whereas for the Li et al study we are not aware of the preoperative level of gross motor disturbance [56], the studies of Langerak et al[54], Golan et al.[57] and Johnson et al.[55] included patients with spastic diplegia who walked with or without assistive devices (though similar to our own cohort) and only the study of Turi et al. [58] mainly consisted in non-ambulators. Furthermore the operative procedures differed between studies and in conclusion it remains unclear to what extent the preoperative levels of gross motor disturbance or the operative procedures might explain differing results with respect to the frequency of spinal abnormalities on long-term after SDR.

In summary data on the long-term outcome of SDR in children with spastic CP are very limited and it remains unclear to what extend SDR influences outcome on long-term. Studies assessing long term outcomes are always subject to many methodological difficulties. For example patients undergoing SDR often require additional orthopedic surgery [45;52;53;59–61] or treatment with botulinum toxin. In general patients with spastic CP undergoing SDR are involved in regular physical therapy programs, which may differ considerably from place to place and from patient to patient. It mostly remains unclear to what extent the SDR itself or the additional treatments mainly influenced the outcome parameters.

Final conclusions, implications and recommendations

This thesis provides information with respect to short and long term outcomes after SDR in children who suffer from spasticity due to different etiologies. The value of preoperative neuroimaging has been assessed in a prospective cohort study. Patients with different patterns on neuroimaging of the brain show differing outcomes with respect to gross motor function. On short term the outcomes with respect to gait performance seems to be dependent on the preoperative level of gross motor disability.

In patients with progressive neurological disease it has been shown that SDR might represent a treatment option for the ease of care and the improvement of sitting comfort. On long term SDR seems to have a positive impact on gait performance, but it remains unclear to what extent SDR improves gross motor function and other domains of the patient's health condition. Spinal deformities seem to be common after SDR on long term. For the clinical application this thesis provides important insights with respect to the selection of patients and to the necessary of follow up examinations.

Clinical Implication

Patients with neurodegenerative disease show a reduction of spasticity, an improvement of sitting comfort and an ease of care after SDR. The progression of the disease might still lead to a functional deterioration and other motor disturbances than spasticity – such as ataxia or diskinesia – cannot be influenced by SDR. In patients who suffer from spasticity due to neurodegenerative disease, who are severely disabled and lost their ambulation, SDR can be recommended as a treatment option for the ease of care. The effect of SDR in ambulatory patients suffering from neurodegerative disease remains unclear and it is unknown to what extent SDR might influence gait performance. Therefore we do not consider SDR as a treatment option for the improvement of gross motor function.

Neuroimaging can be of help in the selection process for SDR and we recommend MRI studies of the brain and – if indicated- of the spinal cord in all patients who are candidates for SDR. Neuroimaging might be necessary for the diagnostic evaluation of the patient with bilateral spastic paresis and specific causes of the spasticity might be detected. Treatment strategies may considerably depend of the etiology of the spasticity. When SDR is considered as a therapeutical option in patients with bilateral spasticity and preserved walking ability, children with different patterns of brain lesions may show different outcomes after SDR with respect to their gross motor skills. Patients with normal MRI findings of the brain might profit considerably from SDR and – if they meet other selection criteria – can be considered as candidates for SDR. In contrast patients with hydrocephalus might not profit from SDR and therefore we do not recommend SDR in patients with hydrocephalus.

In children with bilateral spastic CP who are treated with SDR to improve gross motor function, patients with lesser gross motor impairment – especially those who walk without assistive devices before the operation – seem to profit more from SDR with respect to their gait performance on short term. Though, we can recommend SDR as a treatment option for the improvement of gait performance in patients diag-

nosed with bilateral spastic CP who are classified in GMFCS level I or II (subject to the condition that other selection criteria are given). Children with bilateral spastic CP in GMFCS level III may also profit from SDR with respect to their gait performance. However, in children who walk with assistive devices we recommend a very careful evaluation of muscle force and selective motor control before the operation. Only children with good muscle force in the knee extensors, the hip extensors and the ankle plantar flexors should be defined as candidates for SDR. A treatment trial with botulinum toxin in the most spastic muscle groups and a standardized gait assessment before and after the trial might answer the question whether the reduction of spasticity due to the SDR might contribute to instability and aggravate muscle weakness.

Although it remains unclear to what extend SDR influences the patient's health condition on long term, we still consider SDR as a good therapeutic alternative for the treatment of lower limb spasticity in patients with bilateral spasticity. However, it is essential that patients who undergo SDR are followed on a regular base at least until their adulthood and – if necessary additional treatments are offered. Long-term outcome studies after SDR have shown deformities of the spine in large percentage of patients. Although it cannot be concluded that the spinal deformities are in direct relation with the SDR, in the preoperative evaluation a very thorough clinical and radiological examination of the spine is mandatory. Patients who already show spinal deformities in the preoperative assessment and patients with marked truncal hypotonia who present a risk for spinal deformities on longer term should not undergo SDR. Follow Up assessments should – besides regular assessments by physical therapists and gait assessment – also include a clinical and radiological examination evaluation of the spine.

In patients who undergo SDR to improve their gait performance a regular gait assessment should be performed and compared with the preoperative findings. Three dimensional gait analyses is the gold standard for the assessment of gait abnormalities in patients with spastic CP. However, three dimensional gait analyses is often not available and depends largely on the patient's cooperation. Therefore alternatives, such as the observation of gait on video are performed to document gait abnormalities in children with spastic CP. When three dimensional gait analysis cannot be performed we recommend gait assessment by validated measures that have specifically been developed for this purpose in children with spastic CP. When – besides the overall gait performance – gait kinematics should be measured, we can recommend video screen measurement of the sagittal gait kinematics – especially in the knee joint. However, the observers must be aware that – especially when rotational abnormalities are present – video screen measurement does not necessarily reflect the real position of a single joint.

Recommendations for the Future

In clinical research the randomized controlled trial is the best way to answer question with respect to the efficiency of an intervention. However, in the field of pediatric rehabilitation medicine this study design is often unrealistic for several reasons. The populations which undergo a certain interventions are often not homogeneous and the interventions and their follow-up therapies can be very variable and are not easy to standardize. Study populations can be quite small and it may be very difficult to recruit enough study participants to have sufficient power to answer a research question. Multicenter studies – which would enable larger study groups – are subject to large organizational difficulties and may be very expensive to perform [62]. Other study designs than the RCT therefore might be more suitable to answer "the big questions" in pediatric neurodisability [63]. Furthermore, to assess whether an intervention is successful or not is very much dependent on the choice of the outcome parameter. In rehabilitation medicine interventions are often not simply focused to improve anatomical parts or physiological functions of the body, but may also aim to improve the execution of a task or an action or the involvement in a life situation. Models such as "The International Classification of Functioning, Disability and Health" described how people live with their health condition and have increased the awareness that outcomes after an intervention should focus on different domains of the health condition, which are classified from body, individual and societal perspectives [64]. In patients with cerebral palsy for example a reduction of spasticity does not necessarily mean that gross motor function and functional skills are improved in the same way and the interactions between spasticity, strength and gross motor function are complex [65]. Outcome parameters which are used to assess the effect of an intervention must be chosen with caution. Whenever possible, standardized measures, which have been evaluated for the use in patients with CP and which show good reliability and validity should be used and the outcomes should be assessed in the different domains of the patient's health condition.

With respect to SDR various studies have been performed which assessed the outcome concerning spasticity reduction and gross motor function on short term in children diagnosed with spastic CP [66]. Those studies also included RCTs comparing SDR with postoperative physical therapy compared to physical therapy alone [44;67;68]. The outcomes differed considerably between the studies. Later-on a meta-analysis revealed, that SDR leads to a reduction of spasticity and an improvement of gross motor function in patients with spastic CP undergoing SDR on short term [43].

The comparison of outcomes between different centers is difficult, as preoperative patient characteristics, the operative procedure and the postoperative rehabilitation may not be identical in different centers. Though question arises whether preoperative patient characteristics (such as age, gross motor performance, intellectual abilities) or differences in the operative techniques (namely the amount of rootlets trans-sected by SDR) may explain the differences in outcome between the single RCT's [43]. Only few studies have tried to assess the influence of preoperative patient characteristics on outcome after SDR [27;36;42;45] and their results were inconclusive. Selection criteria for SDR are based on clinical experience and on theoretical considerations rather than on scientific evidence. However, for the selection process it would be crucial to identify the "ideal" patient for SDR and the selection criteria can only be optimized when more details about the influence of preoperative characteristics on outcome are available. Therefore future studies assessing outcome after SDR should not only focus on the question whether SDR improves gross motor function in general, but also try to study the factors which lead to an improved outcome after SDR more in detail. To allow a comparison between studies the selection criteria to undergo SDR should be described explicitly and – if possible – preoperative evaluations should include standardized measures which have been validated for patients with spastic CP and allow a comparison between studies. Furthermore only few studies examined outcome parameters which assessed the participation domain after SDR and none of those allows to draw a conclusion on the causal relation between SDR and the participation issues that have been studied [51;69]. Though future studies should also aim to assess the participation and the quality of live in patients with spastic CP that underwent SDR.

With respect to long term outcome after SDR there is a lack of evidence concerning the efficiency of SDR and future studies are needed to clarify the question, to what extent SDR can lead to improvements of the patient's health condition on long term. Especially for the assessment of long term effects the RCT does not seem to be an ideal way to describe differences between intervention and control group. Already at short term preoperative differences between groups might explain the differing outcomes in different studies. In pediatric rehabilitation medicine RCT's are very difficult to perform because of several reasons and it may very difficult to guarantee homogeneity of patients in clinical studies that compare effects of an intervention between groups. Especially on long term the outcome might be dependent of several covariates. Homogenity between study groups on long term is almost impossible to be maintained. For example it has been repeatedly shown that a large percentage of patients who undergo SDR later are attributed to orthopedic operations [52;53;61] or treatment with botulinum toxin [53]. Orthopedic operation and the treatment with botulinum toxin are known to significantly affect gross motor function in patients with spastic CP and it may remain unclear to what extent the improvements on long term

are mainly resulted by SDR or mainly resulted by other procedures. Furthermore, in clinical practice it is impossible to maintain a standardized and comparable rehabilitation setting in all patients. Differences in the setting of physical therapy (such as the experience of the physical therapist, the methods used in practice, the intensity of the treatment and the cooperation of the child) are inevitable. Though other study protocols than the RCT – such as matched controlled studies might be more suitable to assess long-term outcomes after SDR. Studies assessing long term outcomes after SDR should include larger cohorts and need to address the question to what extent the SDR influenced the patient's health condition in its different domains more in detail. On long-term, spinal deformities seem to be common in patients who underwent SDR, but it remains unclear to what extend spinal deformities are in direct relation with SDR. Though to allow a conclusion whether there is a relationship between SDR and spinal deformities, patients who underwent SDR should be compared with matched controls that did not undergo the operation. The operative techniques of SDR differ between centers or even between single centers. It has not been examined whether different operative techniques (laminectomy, laminotomy, differences in the operated levels, differences in the trans-sected rootlets etc.) have an influence on outcome on long term. Though future studies might address the question whether different operative techniques might lead to differences in outcomes and adverse events – such as spinal instability.

The outcome after SDR in children suffering from spasticity due to other etiologies than spastic CP should be assessed more in detail. However, SDR is very rarely performed in patients suffering from spasticity due to other etiologies than CP and the cooperation of multiple centers would be needed to gain more information on this topic. If possible study groups with patients diagnosed with the same disease should report outcomes in the different areas of the patient's health condition. Patients who suffer from a progressive or a non-progressive neurological disease should be differed from each other. The outcome after SDR in patients suffering from hereditary spastic paraperesis is of special interest, as the clinical presentation in this diagnostic subgroup might be very similar to that of patients suffering from bilateral spastic CP. Patients with spasticity due to injuries or diseases of the spinal cord, patients suffering from spasticity due to anoxic lesions or traumatic brain injury or patients suffering from neurodegenerative diseases would represent other diagnostic subgroups to be examined more in detail. In patients with progressive neurological disease it would be of special interest if the spasticity reoccurs in the long term.

TABLE 1 **Selection Criteria for Selective Dorsal Rhizotomy in Children**

Personal Data
Definite Age[17;21;26;34;37;39;44;52;55;68;70–75]
Clinical Examination
Spastic tone (interfering with motor function)[16–19;21;23;25–27;33;36;37;39;42;44;45;52–55;67–72;74;76–84]
No dystonia/athetosis/ataxia[18;19;23;25;39;45;52;54;67;68;72;74;78;79;81–84]
Good strength[16–18;21;23;25–27;33;34;52–54;68–70;72;74;76;78;82;83]
Good selective motor control[16;18;21;26;27;33;55;70]
No scoliosis[17;26]
No (severe) joint contractures[16;18;21;23;26;27;34;37;39;44;52;53;67;69;71;72;84]
Good to moderate balance[21]
Good trunk control[25;26;44;55;67;70;72;74]
History
History of prematurity[25;26;33;37;55;70;71;73;81]
No chronic medical condition[21]
No (major) skeletal operation[21;23;39;44;52;72;81;84;85]
Gross Motor Function Level
Ambulatory (or potential to walk)*[16;23;25;27;34;53–55;67;73;85]
Cognition
No (severe) mental Retardation[21;23;25;26;36;52;55;67;69;70;72;77;85]
Personal & Environmental factors
Good rehabilitation setting/support[16;21;23;25–27;34;53;55;69;70;77;78;82;83]
Good motivation[21;25;26;39;53;55;70;73]
Additional Investigations
Absence of Basal Ganglia Lesions on MRI[21;37;71]
Evidence of PVLon MRI[33;34]
(Presence of Hydrocephalus on MRI)[27]
No hip dysplasia/joint dislocations[21;67;68;85]
No overweight[21]

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Chapter 9

Summary

Selective dorsal rhizotomy (SDR) is a treatment modality to reduce spasticity which has mainly been evaluated in children who suffer from spastic cerebral palsy (CP). The effect of SDR in paediatric patients who suffer from spasticity due to other etiologies than CP, especially in patients who suffer from progressive spasticity, has not been described previously. Furthermore, the selection of patients for SDR is mainly based on clinical criteria, and other criteria, such as neuroimaging, have not been evaluated in patients undergoing SDR. Although most centers that perform SDR have definite selection criteria, the impact of preoperative clinical findings on outcome has hardly been described. Although there is good evidence that SDR has a positive impact on the reduction of spasticity and the level of functioning in the short term, the long-term effects and adverse events after SDR have only been reported in few studies which were published very recently. The aims of the present thesis were therefore (A) to describe the effects of SDR in patients who suffer from spasticity due to other etiologies than CP, (B) to assess whether neuroimaging studies can help to select patients for SDR, (C) to describe possible relations between the preoperative level of functioning and gait performance after SDR and (D) to describe the long-term effects and adverse events after SDR.

Chapters 1 and 2 comprise the introduction, aims, design and outline of the study.

In **chapter 3** we report the effectiveness of SDR in two patients with progressive spasticity due to neurodegenerative disease. The cases described in chapter 3 showed that spasticity diminished or completely disappeared after SDR, and did not recur. In both patients, caregiving was eased and sitting comfort was improved. However, whereas the spasticity was dramatically and permanently reduced, SDR did not have an influence on other motor disturbances than spasticity, such as ataxia and posture-dependent muscle spasms. We therefore concluded that before deciding to perform SDR in patients who suffer from progressive spasticity due to neurodegenerative disease, other treatment modalities, such as the use of oral antispastic medication or intrathecal baclofen therapy should be considered first. However, when other treatment modalities fail or are contraindicated, SDR could be an alternative to treat spasticity in patients who suffer from spasticity due to progressive neurological disease.

In the study reported in **chapter 4,** we aimed to determine whether there is a relationship between preoperative MRI findings and gross motor function capacities after SDR in patients with bilateral spasticity. We compared the changes in the Gross Motor Function Measure (GMFM) for 19 patients who underwent SDR and for whom preoperative neuroimaging studies were available. The MRIs were classified into three different diagnostic groups: periventricular leucomalacia (PVL, n=10), hydrocephalus (n=2) and normal MRI (n=6). In patients with PVL, the severity of the MRI abnormalities was also scored, using a scoring system which assesses ventricular

size, extension of white matter signal intensity, extension of white matter loss, thinning of the corpus callosum, dimensions of subarachnoidal space, evidence of cysts, and presence of gray matter abnormalities. We compared the changes in the GMFM-66 after SDR between patients who presented with evidence of hydrocephalus, patients who showed evidence of PVL and patients who did not show any MRI abnormalities. Additionally, we correlated the severity of the MRI abnormalities in patients with PVL with the changes in the GMFM-66. After a mean follow-up duration of 5 years and 4 months, we observed the best improvement of gross motor function in patients with normal MRI findings. Patients who showed evidence of PVL in the preoperative MRI had intermediate improvements, whereas the two patients who suffered from hydrocephalus showed no improvement in gross motor function. Whereas the severity of the preoperative MRI abnormalities correlated with the preoperative level of gross motor capacity in patients with PVL, there was no correlation between the severity of the preoperative MRI abnormalities and the improvement after SDR. We concluded that MRI of the brain can provide additional information for the selection of patients for SDR, but that the degree of PVL does not provide information about the degree of improvement in gross motor function after SDR.

In **chapter 5** we assessed the reliability and validity of a custom-made software program that is routinely used at the VU Medical Center to assess gait kinematics in patients with spastic CP. The reasons for performing this study were that we wished to assess gait kinematics before and after SDR, and that for most of the patients preoperative gait analysis included only video documentations and EMG measurements, but not 3-dimensional instrumental gait analysis, which is considered to be the gold standard for the assessment of gait kinematics. A group of 17 patients diagnosed with spastic CP was examined. The patients walked on a 10 m walkway and video recordings were made. Additionally, 3-D instrumental gait analysis was carried out. Two investigators measured 6 different sagittal joint/segment angles (shank, ankle, knee, hip, pelvis and trunk) using a custom-made software package (the MoXie Viewer®, www.smalll.nl). The reproducibility of these measurements was assessed by the intraclass correlation coefficient (ICC), the standard error of measurement (SEM) and the smallest detectable difference (SDD). In addition, the agreement between the video screen measurements and 3-Dinstrumental gait analyses was analyzed by Bland-Altman plots and limits of agreement (LoA). The findings on intra-rater reproducibility showed that the ICC ranged from 0.99 in the shank to 0.58 in the trunk, the SEM from 0.81° in the shank to 5.97° in the trunk and the SDD from 1.80o in the shank to 16.55° in the trunk. With regard to the inter-rater reproducibility, the ICC ranged from 0.99 in the shank to 0.48 in the trunk, the SEM from 0.70° in the shank to 6.78° in the trunk and the SDD from 1.95° in the shank to 18.8° in the trunk. The LoA between the video screen measurements and 3-D gait analyses was best for the knee extension in the stance phase (0.4 +/–13.4°) and poorest for the

ankle dorsiflexion in swing (12.0 +/–14.6°). We concluded that, when performed by the same observer, the video screen measurements allow relevant changes after an intervention to be detected. With respect to the gait changes after SDR, we therefore decided that these gait changes should be measured by the same observer, and we chose to primarily measure changes in the knee kinematics, due to the poorer intra-rater reproducibility for the ankle, pelvis, hip and trunk and the larger LoA between video screen measurements and 3-D instrumental gait analysis in the ankle.

In **chapter 6** we summarize the results of patients with preserved walking ability who have been treated with SDR at the VU Medical Center, Amsterdam, the Netherlands. A gait analysis was performed for 30 children before SDR and 12 to 24 months postoperatively. The subjects walked on a 10 m walkway at comfortable walking speed, and biplanar video was recorded. We measured the sagittal knee angles according to the method described in chapter 4, and administered the Edinburgh Gait Assessment Scale (EGAS) before and after the SDR. In addition, surface EMG was recorded for the rectus femoris (RF) muscle, the vastus lateralis (VL) muscle, the gastrocnemius medialis (GM) muscle, the tibialis anterior (TA) muscle and the semitendinosus (ST) muscle. We compared the gait performance – as assessed by the EGAS – before and after SDR, and correlated the improvement in the EGAS with the age at SDR, the Gross Motor Function Classification Score before SDR and the total EGAS score before SDR. The kinematic measurements of the knee joint were compared before and after SDR. EMG changes after SDR were assessed qualitatively and a quantitatively – describing the total amount of abnormal EMG activity. The EGAS significantly improved after SDR ($p<0.001$) and there were significant improvements in knee angle kinematics ($p<0.001$). The improvement in terms of EGAS scores was significant in independent ambulators ($p<0.001$) as well as in patients who had walked with assistive devices before SDR ($p=0.002$). Improvements were much more pronounced among the independent ambulators and correlated significantly with the preoperative GMFC score ($rho=–0.413$, $p=0.001$), but not with the patient's age at the time of SDR or the preoperative EGAS score. With respect to the EMG activity, only slight changes were observed after SDR. We found that the activity of the GM decreased and a late peak appeared in stance, while the activity of the ST increased in stance. Furthermore, the activity of the RF decreased in swing. In conclusion, we found that SDR improved overall gait performance, especially in patients who had walked without assistive devices before SDR. EMG changes were only slight. Better timing of the GM in stance and reduced activity of RF in swing may have increased knee flexion in swing, and reduced hamstring spasticity may have led to postural instability of the hip.

The study reported in **chapter 7** aimed to assess the long-term outcome and adverse events of SDR in children with spastic CP. We performed a systematic search of the medical literature and summarized the results in a systematic review. The studies included in this review reported results of children with CP who underwent SDR with a follow-up period of at least five years. At least 50% of the subjects were diagnosed with CP or the outcome results had to be documented separately for patients diagnosed with CP. Only articles written in English were included in the review. Several databases were screened (MEDLINE, Web of Science, Embase, PEDro, and the Cochrane library) and the studies meeting the inclusion criteria were scored by two reviewers. The studies were graded with respect to their level of evidence and their quality/conduct. In total, we identified 21 studies that met the inclusion criteria; these included 966 patients. On the whole, their strength and the quality of the evidence were very limited. Only three Level III studies provided a tentative conclusion and it could be assumed that the outcomes were attributable to the SDR. All 18 remaining level IV studies did not allow a conclusion with respect to the causation of SDR and the outcomes 5 years or more after SDR. Two level III studies reported outcomes after 10 and 20 years for the ICF domains of body structure and function. Both reported improvements of gait after SDR. One level III study reported outcomes in the ICF domain of activity; it did not find any differences in the activity domain between children undergoing SDR and a control group that was treated with physical therapy alone. There were no studies reporting outcomes in the ICF domain of participation. Various studies with lower levels of evidence reported spinal abnormalities after SDR. In summary, we found that there is moderate evidence that SDR has a positive long-term influence on the ICF domains of body structure and body function, but that there is no evidence that SDR has an effect on the ICF activity and participation domains.

Chapter 8 presents a general discussion. Methodological issues and aspects of the outcome assessments after SDR are discussed in detail. The possibility of SDR as a treatment option for spasticity in patients diagnosed with other disorders than CP is discussed. The chapter also discusses possible relationships between the preoperative level of functioning and the improvement in terms of gross motor function and gait performance after SDR, as well as the value of preoperative neuroimaging and the long-term effects and adverse events of SDR. The chapter concludes by discussing clinical implications and formulating recommendations for future research.

Chapter 10

Nederlandse Samenvatting

Selectieve dorsale rhizotomie (SDR) is een behandeling voor spasticiteit, die voornamelijk is geëvalueerd bij kinderen met spastische cerebrale parese (CP). Het effect van SDR bij pediatrische patiënten met spasticiteit ten gevolge van een andere etiologie dan CP, en met name bij patiënten met progressieve spasticiteit, is nog niet eerder beschreven. Voorts is de selectie van patiënten voor SDR voornamelijk gebaseerd op klinische criteria, en is de waarde van andere criteria, zoals neuroimaging, bij patiënten die voor SDR in aanmerking komen, nog niet geëvalueerd. Hoewel de meeste centra waar SDR wordt uitgevoerd, duidelijke selectiecriteria hanteren, is er weinig bekend over de relatie tussen preoperatieve klinische bevindingen en de uitkomsten. Hoewel er goede aanwijzingen zijn dat SDR op de korte termijn een gunstige uitwerking heeft op de spasticiteit en het functioneringsniveau, zijn het effect op langere termijn en de ongewenste bijwerkingen pas in enkele zeer recente studies beschreven. Doel van dit promotieonderzoek was daarom: A) het beschrijven van de effecten van SDR bij patiënten met spasticiteit als gevolg van andere etiologie dan CP, B) onderzoeken of neuroimaging nuttig kan zijn bij het selecteren van patiënten voor SDR, C) het beschrijven van mogelijke relaties tussen het preoperatieve functioneringsniveau en het gangbeeld na SDR en D) het beschrijven van de effecten van SDR en de eventuele ongewenste bijwerkingen ervan op de langere termijn.

In **hoofdstuk 1 en 2** wordt een algemene inleiding gegeven, en wordt een kort overzicht gegeven van doel en opzet van het onderzoek.

In **hoofdstuk 3** worden de resultaten van SDR beschreven bij 2 patiënten met progressieve spasticiteit als gevolg van een neurodegeneratieve aandoening. Uit de in dit hoofdstuk beschreven casussen blijkt dat de spasticiteit na de ingreep verminderde of geheel verdween, en niet terugkwam. Beide patiënten hadden na de ingreep minder zorg nodig en hun zitcomfort was verbeterd. Maar hoewel de spasticiteit dus drastisch en permanent was verbeterd, had de SDR geen invloed op andere motorische stoornissen dan spasticiteit – zoals ataxia en houdingsafhankelijke spasmen. Geconcludeerd werd dat alvorens te beslissen om SDR toe te passen bij patiënten met progressieve spasticiteit ten gevolge van neurodegeneratieve aandoeningen, eerst andere behandelmogelijkheden, zoals orale antispastische medicatie of intrathecaal baclofen, dienen te worden overwogen. Indien andere behandelopties niet het gewenste effect hebben of gecontra-indiceerd zijn, kan SDR ook voor patiënten met spasticiteit tengevolge van een progressieve neurologische aandoening een bruikbaar alternatief vormen.

Doel van het in **hoofdstuk 4** besproken onderzoek was om te bepalen of er bij patiënten met bilaterale spasticiteit sprake is van een verband tussen de preoperatieve MRI-bevindingen en de grof-motorische vaardigheden na SDR. Daartoe werden bij 19 patiënten die waren behandeld met SDR en van wie preoperatieve neuroimaging gegevens beschikbaar waren, de veranderingen op de Gross Motor Function Measure (GMFM) vergeleken. De MRI-beelden werden onderverdeeld in drie diagnostische groepen: periventriculaire leukomalacie (PVL, n=10), hydrocephalus (n=2) en normale MRI (n=6). Bij de patiënten met PVL werd tevens de ernst van de MRI-afwijkingen bepaald met behulp van een scoringssysteem op basis van ventrikelgrootte, extensie van de signaalintensiteit van de witte stof, verlies van witte stof, dunner worden van het corpus callosum, afmetingen van de subarachnoïdale ruimte, aanwijzingen voor de aanwezigheid van cysten en aanwezigheid van grijze stof afwijkingen. De veranderingen op de GMFM-66 na SDR werden vergeleken bij patiënten met verschijnselen van hydrocephalus, patiënten met verschijnselen van PVL en patiënten zonder afwijkingen op de MRI. Voorts werd bij de patiënten met PVL de ernst van de MRI-afwijkingen gecorreleerd aan de veranderingen op de GMFM-66. Na een gemiddelde follow-up termijn van 5 jaar en 4 maanden werd de grootste verbetering van de grove motoriek gevonden bij de patiënten met normale MRI-bevindingen. Bij de patiënten met verschijnselen van PVL op de preoperatieve MRI was er sprake van matige verbetering, terwijl bij de twee patiënten met hydrocephalus geen verbetering in de grove motoriek werd geconstateerd. Bij de patiënten met PVL correleerde de ernst van de preoperatieve MRI-afwijkingen wel met het preoperatieve niveau van de grove motoriek, maar er werd geen correlatie gevonden tussen de ernst van de preoperatieve MRI-afwijkingen en de na SDR opgetreden verbetering. Wij concluderen dat een MRI van de hersenen aanvullende informatie kan leveren bij het selecteren van patiënten voor SDR, maar dat de ernst van de PVL geen informatie verschaft over de te verwachten verbetering van de grove motoriek na SDR.

Doel van het in **hoofdstuk 5** besproken onderzoek was het bepalen van de betrouwbaarheid en validiteit van een speciaal ontworpen softwareprogramma dat bij het VU Medisch Centrum in de dagelijkse praktijk wordt gebruikt voor de beoordeling van gangbeeld-kinematica bij patiënten met spastische CP. De aanleiding tot het onderzoek was dat we de gangbeeld-kinematica wilden beoordelen voor en na de toepassing van SDR. Bij de meeste patiënten omvat de preoperatieve gangbeeldanalyse alleen video-opnamen en EMG-metingen, en geen driedimensionale instrumentele gangbeeldanalyse, die geldt als de gouden standaard voor het beoordelen van de gangbeeld-kinematica. Het onderzoek omvatte een groep van 17 patiënten met de diagnose spastische CP. De patiënten liepen op een looptraject van 10 meter, waarbij video-opnamen werden gemaakt. Daarnaast werd een 3D instrumentele gangbeeldanalyse uitgevoerd, waarbij twee onderzoekers zes verschillende sagittale gewrichts/segmenthoeken (onderbeen, enkel, knie, heup, bekken en romp) bepaalden met be-

hulp van een speciaal ontwikkeld softwarepakket (MoXie Viewer®, www.smalll.nl). De reproduceerbaarheid van deze metingen werd bepaald met behulp van de intra-class correlatie coëfficiënt (ICC), de standaardfout (standard error of measurement = SEM) en het kleinste detecteerbare verschil (smallest detectable difference = SDD). De mate van overeenstemming tussen de video-metingen en de 3D instrumentele gangbeeldanalyse werd beoordeeld aan de hand van Bland-Altman plots en de grenzen van overeenstemming (Level of Agreement = LoA). Uit de bepaling van de reproduceerbaarheid bleek dat de ICC varieerde van 0,99 voor het scheenbeen tot 0,58 voor de romp, de SEM van 0,81° voor het scheenbeen tot 5,97° voor de romp en de SDD van 1,80o voor het scheenbeen tot 16,55° voor de romp. Uit de analyse van de inter-waarnemer reproduceerbaarheid bleek dat de ICC varieerde van 0,99 voor het scheenbeen tot 0,48 voor de romp, de SEM van 0,70° voor het scheenbeen tot 6,78° voor de romp, en de SDD van 1,95° voor het scheenbeen tot 18,8° voor de romp. De meest gunstige LoA tussen de videometingen en de 3D gangbeeldanalyse werd gevonden voor de knie-extensie in de standfase (0,4 +/–13,4°) en de minst gunstige voor de enkel-dorsiflexie in de zwaaifase (12,0 +/–14,6°). Wij concludeerden dat het met videometingen mogelijk is relevante veranderingen na een interventie te bepalen, mits ze worden uitgevoerd door dezelfde waarnemer. Daarom werd besloten dat de veranderingen in het gangbeeld voor en na de SDR door dezelfde waarnemer moesten worden bepaald en dat we primair de veranderingen in de kinematica van de knie zouden bepalen, gezien de minder goede inter-waarnemer reproduceerbaarheid van de metingen voor enkel, bekken, heup en romp, en de hogere LoA tussen de videometingen en de 3D instrumentele gangbeeldanalyse voor de enkel.

In **hoofdstuk 6** worden de resultaten samengevat van patiënten die hun loopvermogen hadden behouden en die in het VU Medisch Centrum in Amsterdam werden behandeld met SDR. Een gangbeeldanalyse werd uitgevoerd bij 30 kinderen, voorafgaand aan de ingreep en 12 en 24 maanden erna. Terwijl de kinderen met comfortabele snelheid liepen over een traject van 10 meter, werd een biplanaire video opgenomen. De sagittale gewrichtshoeken van de knie werden gemeten met behulp van de in hoofdstuk 4 besproken methode. Daarnaast werd voor en na de SDR de Edinburgh Gait Assessment Scale (EGAS) toegepast, en werd een oppervlakte-EMG gemaakt van de volgende spieren: rectus femoris (RF), vastus lateralis (VL), gastrocnemius medialis (GM), tibialis anterior (TA) en semitendinosus (ST). Het gangbeeld – zoals vastgesteld met de EGAS – werd vergeleken voor en na de SDR. De verbetering op de EGAS werd gecorreleerd aan de leeftijd ten tijde van de SDR, de score op de Gross Motor Function Classification (GMFC) vóór de SDR en de EGAS totaalscore vóór de SDR. De kinematische metingen aan het kniegewricht voor en na de SDR werden vergeleken. De veranderingen in de EMG na de SDR werden kwalitatief en kwantitatief beoordeeld – ter beschrijving van de totale omvang van de afwijkende EMG-activiteit. De EGAS-score bleek na de SDR significant te zijn verbeterd

($p<0,001$) en ook werden significante verbeteringen gevonden in de kinematica van het hoekgewricht van de knie ($p<0,001$). De verbetering van de EGAS-scores was significant bij de kinderen die zelfstandig konden lopen ($p<0,001$) evenals bij degenen die vóór de SDR loophulpmiddelen gebruikten ($p=0,002$). Bij degenen die zelfstandig konden lopen, waren de verbeteringen veel geprononceerder, en correleerden significant met het preoperatieve GMFCS-score (rho=–0,413, p=0,001), maar niet met de leeftijd waarop de SDR werd uitgevoerd, noch met de preoperatieve EGAS-score. Wat betreft de EMG-activiteit werden na de SDR slechts geringe veranderingen geconstateerd. De activiteit van de GM nam af, en er verscheen een late piek in de standfase, terwijl de activiteit van de ST in de standfase toenam. Voorts nam de activiteit van de RF tijdens de zwaaifase af. De conclusie van het onderzoek luidde dat SDR een verbetering teweegbracht in het algehele gangbeeld, vooral bij patiënten die vóór de ingreep geen loophulpmiddelen gebruikten. In het EMG traden slechts geringe veranderingen op. De flexie van de knie tijdens de zwaaifase was wellicht verbeterd als gevolg van een betere timing van de GM in de standfase en een geringere activiteit van de RF tijdens de zwaaifase. Een afname van de spasticiteit in de hamstrings kan hebben geleid tot posturale instabiliteit van de heup.

Doel van het in **hoofdstuk 7** beschreven onderzoek was het beoordelen van de uitkomsten en de eventuele ongewenste bijwerkingen van SDR bij kinderen met spastische CP op langere termijn. Dit werd gedaan via een systematische review op basis van een systematische literatuurstudie. De in de review meegenomen studies hadden betrekking op de resultaten bij kinderen met CP die SDR ondergingen, en die minstens vijf jaar waren gevolgd. Minstens 50% van de deelnemers had de diagnose CP, of de resultaten van degenen met CP werden afzonderlijk gerapporteerd. Alleen artikelen in het Engels werden meegenomen in de review. Artikelen werden gezocht in verschillende databases (MEDLINE, Web of Science, Embase, PEDro en de Cochrane library) en de studies die aan de selectiecriteria voldeden, werden op kwaliteit beoordeeld door twee reviewers. De studies werden beoordeeld op basis van het niveau van de evidentie en de kwaliteit en uitvoering. In totaal werden 21 studies gevonden die aan de criteria voldeden; deze hadden betrekking op in totaal 966 patiënten. Over het algemeen waren de sterkte en de kwaliteit van de evidentie zeer beperkt. In slechts 3 studies van niveau III werd een voorzichtige conclusie gepresenteerd waarbij kon worden aangenomen dat de bereikte resultaten toe te schrijven waren aan de SDR. Bij de overige 18 studies van niveau IV kon geen conclusie worden getrokken over een causaal verband tussen de SDR en de uitkomsten vijf of meer jaar na de ingreep. Bij twee studies van niveau III werden uitkomsten gemeld na 10 en 20 jaar op basis van de ICF-domeinen functies en anatomische eigenschappen. In beide studies werd een verbetering van het gangbeeld gevonden na de SDR. Bij één studie van niveau III werden uitkomsten gerapporteerd voor het domein “activiteiten”; voor dit domein werden er geen verschillen gevonden tussen de kinderen die met SDR waren

behandeld en een controlegroep die alleen fysiotherapie kreeg. Bij geen van de studies werden uitkomsten gemeld op het ICF-domein “participatie”, maar bij verschillende studies met een lager evidentieniveau werd melding gemaakt van afwijkingen in de ruggengraat na SDR. Samenvattend kan worden gesteld dat er matig sterk bewijs is dat SDR op de langere termijn een gunstige invloed heeft op de ICF-domeinen functies en anatomische eigenschappen, maar dat er geen bewijs is dat de ingreep ook invloed heeft op de domeinen activiteiten en participatie.

In **hoofdstuk 8** wordt een algemene discussie van het onderzoek gepresenteerd, waarbij methodologische kwesties en bepaalde aspecten van het beoordelen van de uitkomsten na SDR in detail worden besproken. Voorts worden de mogelijkheden geëvalueerd van het gebruik van SDR ter behandeling van patiënten met andere diagnoses dan spastische CP. Tevens worden mogelijke verbanden besproken tussen het preoperatieve functioneren van de patiënt en de verbetering in de grove motoriek en het gangbeeld na SDR, alsmede de waarde van preoperatieve neuroimaging en de effecten en ongewenste bijwerkingen van SDR op langere termijn. Ten slotte worden de klinische implicaties van de onderzoeksresultaten besproken en worden aanbevelingen gedaan voor verder onderzoek.

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An meinen Lehrer

Ich war nicht einer Deiner guten Jungen.
An meinem Jugendtrotz ist mancher Rat
Und manches wohlgedachte Wort zersprungen.
Nun sieht der Mann, was einst der Knabe tat.

Doch hast Du, alter Meister, nicht vergebens
An meinem Bau geformt und Dich gemüht.
Du hast die besten Werte meines Lebens
Mit heißen Worten mir ins Herz geglüht.

Verzeih, wenn ich das Alte nicht bereue.
Ich will mich heut wie einst vor Dir nicht bücken.
Doch möcht ich Dir für Deine Lehrertreue
nur einmal dankbar, stumm die Hände drücken.
Joachim Ringelnatz

In October 2005 Prof. Maja Steinlin suggested to me to start a career in pediatric rehabilitation medicine. It was not what I have originally been planning for my professional future. Maja knew it better. As soon as I got familiar with the idea, there was no way back. She proposed a training abroad. My wife Silvia and I have always liked the idea to spend one or two years in a foreign country. We did not know much about the Netherlands. However, both of us thought that it would be an interesting country to go. Though, I asked Maja whether she would know Dutch colleagues working in the field. It was through Prof. Linda de Vries that we first got in contact with Prof. Jules Becher. Jules invited me to a job interview in Amsterdam with the option to do a fellowship in pediatric rehabilitation medicine. When preparing for the interview – amongst others – I consulted "Pubmed" to look for Jules' latest publications. I read an article that had recently been published by Dr. Petra van Schie, Dr. Jeroen Vermeulen, Dr. Willem van Ouwerkerk, Prof. Gert Kwakkel and Prof. Jules Becher and described the outcomes one year after SDR in children diagnosed with bilateral spastic cerebral palsy. I did not know much about the method and at that time SDR was not performed in Switzerland. The first months in the Netherlands were not easy. I had to learn a new language, get to know a new culture and gently started to gain ground in the field of pediatric rehabilitation medicine. As I was also interested to participate in research I spoke with Jules Becher and Jeroen

Vermeulen and we soon started to set up the first plans for a study that aimed to assess preoperative neuroimaging findings in children that have undergone SDR at the VU University Medical Centre. What started as a conversation in Majas office became a passion and what started as an idea became a book. Six years later my thesis is ready for printing. Of course this was not possible without the help and the support of many colleagues, friends and family.

First of all I would like to thank Prof. Jules Becher, my former supervisor and promotor. Not only did you teach me the principles of pediatric rehabilitation medicine. You are definitely an inspiring person. Your work with handicapped children is always passionate and respectful – in everyday's live and also in very busy days. It was surely not easy for you to work with me in the beginning. I rarely spoke Dutch, did not know the medical system in the Netherlands and was a beginner in rehabilitation medicine. You took the time (which I know is a very precious property and always scarce), explained, answered to my questions and corrected me when it was necessary (which I did not always like). In that way I learned to independently care for children with disabilities and now I even pass your knowledge to others. You were insisting that everyone in the department uniquely talks Dutch to me. In that way – thanks to you – it became possible to understand, talk and write the Dutch language within a few months. You were a very creative, a very patient but also a demanding teacher. As a pediatrician I sometimes felt somewhat lost in the department. However, I only later realized that thinking differently is a part of the formation. I learned to think like a physiatrist and still remained a pediatrician in my heart. I know that you first had some skepticism when I intended to participate in your research group besides the clinical work. Anyhow, you gave me the chance and supported my plans. You took the time and listened to my ideas. You taught me how to put up research plans in a realistic way. I now realize how important this is. I always had enough ideas, but tended to do various thinks at once. Thanks to you I advanced step by step and finally finished my thesis without haste. You supported me with all necessary information and gave me the opportunity to build up my own scientific network. You accompanied me carefully, gave me the liberty to bring own ideas into my work but helped me to avoid major mistakes. Dear Jules, you have been a great teacher!

Prof. Maja Steinlin is the person that probably influenced my professional live most of all. Maja, you realized my passion for the field of rehabilitation medicine even before me. You directed me towards this way, but let me do my choice on my own. You supported me when I planned my fellowship. Without your help I would not even have dreamt about a PhD thesis in the Netherlands. When I was abroad you always supported me through distance. An E-Mail or a phone call is sometimes magic when you are abroad. You knew that! You always gave me the feeling that – when

leaving the Netherlands – I could expect a warm welcome in Berne. Your visit to Amsterdam was very important to me and I was somewhat proud to show you what I have been learning. After my return to Switzerland I could continue my clinical work. I had the possibility to organize my own consultancy and to bring in my knowledge – also when it sometimes differed from other concepts. You have supported me – clinically and scientifically. Even when far away you have always taken my concerns seriously and helped me to find creative solutions in complex situations. Your knowledge in the field of pediatric neurology is boundless. I admire you for your strength of purpose and your spirit of enterprise. You are demanding – but you provide the support that is necessary to bring a good performance. You are a trustworthy and supporting mentor, an excellent doctor and a wonderful person. Thank you.

I would also like to thank to Dr. Jeroen Vermeulen. Jeroen, you have believed in our projects from the very beginning and always gave me the support that I needed. You took the time, sat down with me and discussed the projects in detail. We have gathered ideas, prepared study protocols, discussed many articles, looked at MRI's, scored gait patterns, prepared statistic calculations, evaluated results and wrote articles together. I remember the many Nespresso capsules that I consumed in your office. The warm hospitality, the open and tolerant, but also very productive and goal oriented atmosphere impressed me. You motivated me to go on with my research and finally bring all my articles together in a PhD thesis. You were never unfriendly and always patient. You also came to visit us in Berne. We all have kept your visit in very good memory. When I needed to travel to the Netherlands you and your family gave me a place to stay. I still remember the very cold winter when we went ice skating. Besides scientific considerations there was always time to share other interests – such as Jazz Music or winter sports. I am still hoping that we will be able to go skiing in the Bernese Alps once in a while. Jeroen, I am very grateful to you.

When I came to the Netherlands I did not know much about gait analysis. The gait laboratory in the VUMC was indeed the first one I visited. Besides Jules it was Dr. Jaap Harlaar that fascinated me for the clinical use of gait analysis. This started from the very beginning. Every Monday afternoon we had the "Gengbeeldbespreking" – a great opportunity for clinicians and scientists to share their experience. I still remember when I had to present my first case. Not easy, as I needed to do a presentation in Dutch AND needed to talk about gait analysis, which both were new for me. Anyhow, you never gave me the feeling that I did it wrong (even when it was obviously the case). Your comments were always constructive. You looked for feasible solutions and presented complex matters in an easy and understandable way. During the work on my thesis you have given me a lot of technical support. You have prepared the software that I needed for my assessments and even provided a written

instruction, which was very useful and understandable. I could just go on with the measurements without having ANY technological interruption. When preparing the manuscripts and revisions your comments were very constructive and timely. Thank you very much.

When I had the idea to assess the reproducibility of the measurements that we made in daily routine with the "Moxie-Viewer", Dr. Caroline Doorenbosch and Dr. Marjolein van der Krogt helped me from the beginning. Caroline developed the software that we needed for this work within a few days. Marjolein gave us the possibility to use gait studies that she had made before for her own thesis. Though Dr. Petra van Kampen and me could go on and assess the gait kinematics with the "Moxie-Viewer" without technical delays and were not dependent on gait assessments that had to be performed first. Later-on when I needed to do the statistical calculations, Dr. Merel Brehm offered me her help and showed me how to assess the repeatability of measurements. You all have been helpful in the preparation and the revision of the manuscript. Caroline, Marjolein, Petra and Merel – I am very thankful for your co-operation.

Dr. Petra van Schie had published an article with respect to SDR even before I have come to the Netherlands. Though she knew a lot about this matter and had collected a large amount of literature. When I started to show more interest about SDR Petra supplied me with many articles and gave me a lot of very useful information. At that time Petra organized the follow up assessments after SDR. These included the Gait Analysis and the GMFM measurements – both of which I included in my thesis. Petra, you were very helpful in gathering all the data I needed for this work. Furthermore, you helped us in the data interpretation and the preparation of the manuscripts. Many thanks!

Despite all theoretical and scientific considerations medicine remains a practical profession. A thesis dealing with outcomes after an operative procedure could never be possible without a surgeon. Dr. Willem van Ouwerkerk has operated all of the children that have been included in this thesis from the very beginning on. Pim, it was great to be present in the operation theatre while you were operating. I was impressed how concentrated, calm, humorous and attentive to others you were, while performing a surgical top performance. You are indeed a great surgeon! Thank you very much!

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Only through the daily work of clinicians medical science becomes possible. In a hospital setting patient care remains our principle duty. The fact that physicians have a large workload is probably a universal phenomenon and it is a matter of fact that one person cannot do various things at the same time. When working on scientific projects sometimes our colleagues need take over the patient care. I am gratefully thankful to all my colleagues in Amsterdam and in Berne for that!

Working on my thesis would never have been possible without the help and support of many friends – in the Netherlands and in Switzerland. I am indeed a very lucky person to have as many good friends! All of you have always believed in me and given me a lot of support support in many ways. I am gratefully thankful for that.

I want to pronounce a very special thank to all my family – my mother, my sister, my mother in law, my brothers and sisters in law and all nieces and nephews. I knew that I always could count on your support even when being far away from you. Many times you travelled to the Netherlands to be with us and helped us to get everything settled . You spent many days in our house in Haarlem and I will always keep those days in a very good memory. You never stopped to offer your support and now – being back in Switzerland you are always present when I need your help. I am very thankful to all of you.

When I decided to accept the offer of a fellowship abroad, my wife Silvia supported me and we took the decision to go the Netherlands together. It was surely not easy for her. Especially during the first year it needed a lot of time and effort to settle down in a foreign country, to organize the daily living and to build up a new social network. We had very difficult moments at that time. However, in retrospective we are both very glad that we have spent those years abroad. When later-on we did not know if would be possible to stay for a second year and we did not have the finances at the time, Silvia motivated me to look for financial support, to do work in a scientific base additionally and believed in me. Without her support I would not have taken this decision – a decision with consequences for me and my family. I spent many hours writing – mostly not during my regular working hours. I could not spend this time with my wife and my children Béla and Luna. However, Silvia gave me the liberty and motivation to go on and finish this thesis. She then designed the cover and processed the layout. Silvia, I am very, very grateful for your support. Béla and Luna, I thank you very much for your patience and your comprehension.

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