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**The Long-term Sequelae of Selective Dorsal Rhizotomy in  
Patients with Spastic Diplegia**

A Thesis Presented for the Degree of

**Doctor of Philosophy**

at the

University of Cape Town

UCT/MRC Medical Imaging Research Unit,

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# **DECLARATION**

I, Nelleke Gertrude Langerak, do hereby declare this thesis is based on five journal articles: three have been published (Chapters 2, 3 and 4), one has been accepted for publication (Chapter 5), and one is currently being reviewed (Chapter 6). To meet the stylistic requirements of a thesis, the format of the published and submitted papers has been adjusted accordingly and abbreviations of units and terms standardised throughout. Below are the articles, together with a statement of my contribution to these multi-authored papers:

Chapter 2. “Selective dorsal rhizotomy: long-term experience from Cape Town” Langerak NG, Lamberts RP, Fieggen AG, Peter JC, Peacock WJ, Vaughan CL. *Child’s Nervous System*. 2007 Sep;23(9):1003-1006. I drafted a paper based on my own experiences as part of the 20 year follow-up study which was accomplished in 2005 (see Chapter 3 and 4) and summarised the studies published by the Cape Town research group. Together with my collaborators I finalised the manuscript, which was submitted and published in *Child’s Nervous System* in 2007.

Chapter 3. “A prospective gait analysis study in patients with diplegic cerebral palsy 20 years after selective dorsal rhizotomy” Langerak NG, Lamberts RP, Fieggen AG, Peter JC, van der Merwe L, Peacock WJ, Vaughan CL. *Journal of Neurosurgery: Pediatrics*. 2008 Mar;1(3):180-186. I tracked down 100% of the cohort (14 patients: 6 women, 8 men; mean age, 27 years; range, 22–33years) who participated in the original follow-up studies, which included an extended detective search; all these patients are now adults and their contact details were no longer held by Red Cross Children’s Hospital where they were all originally treated. By July of 2005, I completed a road trip around South Africa (Cape Town to Johannesburg to Durban to Port Elizabeth and back to Cape Town) where I, with the assistance of another physiotherapist (Robert P Lamberts), performed both gait analyses and clinical evaluations of their neuromuscular function (Chapter 4). I was then able to re-analyse the gait data of all the original follow-up studies and completed statistical analyses jointly with a statistician at the Medical Research Council (Dr. Lize van der Merwe). I drafted the manuscript, discussed input from my collaborators, edited the paper and

submitted it to the *Journal of Neurosurgery: Pediatrics*, which accepted and published the paper at the beginning of 2008.

Chapter 4. “Functional status of patients with cerebral palsy according to the international classification of functioning, disability and health model: a 20 year follow-up study after selective dorsal rhizotomy” Langerak NG, Lamberts RP, Fieggan AG, Peter JC, Peacock WJ, Vaughan CL. *Archives of Physical Medicine and Rehabilitation*. 2009 Jun;90(6): 944-1003. As described above (see Chapter 3) I tracked down 100% of the cohort (14 patients) who participated in the original follow-up studies. Based on the clinical assessments and captured videos I analysed all data. Thereafter I finalised the statistical analyses and wrote the manuscript, which was reviewed by my collaborators. After adding their suggested changes, I submitted the paper to *Archives of Physical Medicine and Rehabilitation* which accepted the paper and published it in June 2009.

Chapter 5. “Incidence of spinal abnormalities in patients with spastic diplegia 17 to 26 years after selective dorsal rhizotomy” Langerak NG, Vaughan CL, Hoffman EB, Figaji AA, Fieggan AG, Peter JC. *Child’s Nervous System* 2009 Dec;25(12): 1592-1602. For the second phase of my PhD I was able to track down 32 patients with cerebral palsy who received SDR between 1981 and 1991 (13 women, 19 men; mean age 28 years and range 21-44 years). I was responsible for data collection and together with a neurosurgeon (Professor Jonathan C Peter) we analysed X-rays of 31 patients with cerebral palsy who were able to participate in this study. In addition, I did an in depth search of the archives to retrieve the X-ray data of patients who participated in the earlier spinal studies (Peter *et al.*, 1990; 1993) and did the statistical analyses regarding this follow-up. In addition, we obtained magnetic resonance images (MRI) of the spine, and additional information regarding back pain and clinical assessments, which after analysis were presented as descriptive data. I completed this paper, discussed the draft with my collaborators and, after implementing all their suggested changes, it was submitted to *Child’s Nervous System*. The journal accepted the paper in August and will be published in December 2009.

Chapter 6. "Level of activity and participation in adults with spastic diplegia 17 to 26 years after selective dorsal rhizotomy" Langerak NG, Hillier SL, Verkoeijen PPJL, Peter JC, Fieggan AG, Vaughan CL. *Disability and Rehabilitation* (2009). In review. I requested the Life-Habit questionnaire, became experienced in its use and trained another physiotherapist (Dr. Susan L Hillier) to assist with collection of this data in a total 31 patients (see Chapter 5 for more patient details). In addition, I completed the Functional Mobility Scale (FMS) and used a semi-structured interview to gather patients' characteristics and long-term experiences after SDR. I managed the data, analysed this and completed statistical analyses with the support of a statistician (Dr. Peter P.J.L. Verkoeijen). Thereafter, I wrote the first draft, which was discussed with my collaborators and the final version was submitted to *Disability and Rehabilitation* and a review process started in August 2009.

Neither the substance nor any part of this thesis has been submitted in the past, or is being, or is to be submitted for a degree in this or any other University.

I hereby grant the University of Cape Town free licence to reproduce the above thesis in whole or in part, for the purpose of research.

This thesis is presented for examination in fulfillment of the requirements for the degree of Doctor of Philosophy.

Signature: 

Signed by candidate
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Date: 15<sup>th</sup> August 2009

University of Cape Town

# **ABSTRACT**

Cerebral palsy (CP) is known as a leading cause of disability in childhood. About 80% of patients with CP are diagnosed with the spastic type. Since spasticity leads to secondary abnormalities, it is important to treat spasticity with insight in the early childhood years to prevent secondary problems during a person's lifespan.

The neurosurgical procedure known as selective dorsal rhizotomy (SDR) has particular appeal as it reduces lower extremity spasticity by transecting a percentage of lumbar rootlets, which interrupts the abnormal reflex arc at the spinal cord level that maintains spasticity. This procedure was first described in humans in the early 1900's, but had limited acceptance until it was refined and reintroduced by Professor Warwick Peacock at the Red Cross Children's Hospital, Cape Town, in the early 1980's. A large number of studies have demonstrated the benefits of this neurosurgical procedure. However, these conclusions were based on assessments performed within the first few years after surgery. Therefore, the question remains: what will happen when these patients with CP become adults?

As the manifestation of CP and spasticity is complex, the International Classification of Functioning, Disability and Health (ICF) model, which was developed by the World Health Organisation, has proven to be useful to establish an understanding of the outcomes of SDR. This ICF-model provides a biopsychosocial framework to identify and classify abnormalities across two components: (i) *Body Function & Structure*; and (ii) *Activities and Participation*.

The aim of this doctoral thesis was to investigate the long-term sequelae of SDR in patients with spastic diplegia seen in the context of the broad spectrum of the ICF-model: *Body Function & Structure, Activities and Participation*.

This thesis is based on earlier experience with SDR in Cape Town and consisted of two main research projects. Project I included 20 year follow-up studies after SDR in 14 children with spastic diplegia who received SDR in 1985. The focus of these studies was on gait analysis and also on neuromuscular and functional outcomes.

Project II was based on 32 children who underwent SDR between 1981 and 1991 and were studied in 2008 (17 - 26 years after surgery). They received X-rays and MRI-scans of their spine, were clinically assessed by a neurosurgeon and physiotherapist, completed a general questionnaire, and also a questionnaire related to back pain and life-habits.

More than 15 years after SDR positive outcomes were still demonstrated with regards to the ICF-model component *Body Function & Structure*. Despite the small sample sizes of our study cohorts, it was apparent that the patients with spastic diplegia gained sustained benefits with regards to muscle tone, motor control and muscle strength as demonstrated by improvements in their gait patterns. Disturbed pin-prick sensation, reduced reflexes, some spinal abnormalities and an increase in the incidence of spinal deformities were seen. However, no specific concerns regarding spinal abnormalities were reported. Low back pain was indicated, but this did not lead to disability in daily activities for the majority of patients.

Level of *Activity and Participation* of the patients was relatively good, with most of them being independent and experiencing no problems in accomplishing various life habits. Mobility was seen as the life habit which resulted in most problems, despite the fact that after SDR positive changes in Gross Motor Function Classification System (GMFCS) levels were seen and the majority walked independently. In the long-term, the majority of the patients had positive feelings about the SDR operation, but they reported a lack of care with the transition into adulthood.

We would like to emphasize not only the importance of strict selection criteria, but also the ongoing need for long-term follow-ups evaluations. Further studies are necessary to be able to make more definitive conclusions, to explore mobility outcome measures over a lifespan, to understand the relationship with contextual factors, and to evaluate conformity with other countries.

# Chapter 1

Introduction

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## CEREBRAL PALSY

### **History**

In the mid 1800s cerebral palsy (CP), a term for the condition of reduced movement ('palsy') due to damage to the developing brain ('cerebral'), was in the first place called 'Little's Disease'. It was William J. Little who first described cerebral paralysis. He related musculoskeletal problems with obstetrical complications of birth resulting in lack of oxygen to the baby's brain (Little, 1843; Little, 1862). Thereafter, William Osler (1889) used the plural term cerebral palsies and documented 151 cases. He emphasized the correlation between pathology and clinical findings, and classified the cerebral palsies based on distribution of neuroanatomical pathology into infantile hemiplegia, bilateral spastic hemiplegia (or diplegia) and paraplegia. This was in contrast with the ideas of Sigmund Freud (1893), who made a great contribution in the field of CP after Osler. He recognized the differences in clinical manifestation in patients with similar neuropathology, and suggested to include this in the classification of the patients. He also had different ideas about the aetiology of CP, in which he challenged Little's assumption that CP was caused by perinatal asphyxia. He believed that the brain damage as seen in CP could be caused by multiple factors, which were predisposing congenital, perinatal or post-natal factors. The opinion of Freud regarding CP was advanced for that time. More than a century later, his ideas are still part of our current CP definition, classification and aetiology (Kavcic and Vodusek, 2005).

### **Definition and Classification**

During the last century, especially the last decade, there has been increased attention for CP (Kuban and Leviton, 1994; Jones *et al.*, 2007; Nelson, 2008; Koman *et al.*, 2004; Krigger, 2006; Rosenbaum, 2003; Russman and Ashwal, 2004), and many discussions have taken place regarding aetiology and a correct and most appropriate definition and classification of CP has emerged (Shapiro, 2004; Shevell and Bodensteiner, 2004; Armstrong, 2007). As a result of a meeting of international experts in the field of CP, the proposed definition of Martin Bax *et al.* (2005) was revised, and an Executive Committee generated a Report of the Definition and Classification of Cerebral Palsy, in April 2006 (Rosenbaum *et al.*, 2007). This report includes the following new definition of CP:

*“Cerebral palsy (CP) describes a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to nonprogressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication, and behaviour, by epilepsy, and by secondary musculoskeletal problems.”*

This definition of CP combines different clinical presentations with related activity limitations, which refers to a wide variety of patients. Therefore further classification of CP was proposed by the Executive Committee based on the following four components:

- (i) Motor abnormalities, e.g. spasticity, ataxia or dystonia, and Gross Motor Function Classification System (GMFCS) levels.
- (ii) Accompanying impairments, e.g. epilepsy, hearing and vision problems.
- (iii) Anatomical and neuro-imaging findings, e.g. uni- and bilateral motor involvements and ventricular enlargement or brain anomaly.
- (iv) Causation and timing, e.g. postnatal meningitis.

This classification system should be useful in understanding the condition and for support in decision making regarding treatment. However, the Executive Committee acknowledged that not all clinicians are able to complete this classification for each patient with CP currently. Besides the fact that not everybody is able to undertake neuro-images of the patients (iii), it is also not realistic to think that clinicians are able to give a “clear-cut categorization by cause” (iv).

### ***Aetiology***

In regards to the aetiology of CP, great developments in technology and research resulted in more knowledge. However, as Nelson (2008) mentioned in her recently published review article, there is not one clear causal pathway. More commonly it seems that CP is a result of multiple risk factors (as already hypothesized by Freud a century ago), which include: birth asphyxia; prematurity; atypical intrauterine growth; infection, inflammation, and maternal fever in labour; perinatal ischaemic stroke;

congenital anomalies; multiple gestation; placental pathology; genetics; neonatal encephalopathy; or even other factors which have not been pursued in the literature (e.g. polycystic ovary syndrome) (Nelson, 2008).

### ***Prevention and Prevalence***

Despite the distinctive knowledge developed over the last few decades, and thereby the reduced exposure to associated risk factors, there has not been significant decrease in the number of newborn children with CP. Research groups have claimed that the prevalence of CP in developed countries has not decreased since the 1970s (Clark and Hankins, 2003) or has even risen from 1.5 to 2.0-2.5 per 1000 live births in the 1990s (Odding *et al.*, 2006; Stanley *et al.*, 2000a). These unexpected findings led to a number of discussions between researchers and clinicians. In 2009 Baxter wrote an editorial entitled “Preventing cerebral palsy: hidden improvements”, wherein he emphasized that although the prevalence of CP remains steady, worthwhile gains in prevention have been achieved. The number of newborn CP patients might still be about 2 per 1000, but there is decrease in the number of some specific causes and there is a decrease in severity (Baxter, 2009).

### ***Aging and Secondary Abnormalities***

Over last few decades it has been suggested that the survival of patients with CP has increased (Strauss *et al.*, 2007; Krigger, 2006; Kuban and Leviton, 1994). However, Strauss *et al.* (2008) recently clarified and updated the outcomes of life expectancy, and expressed some doubts whether the survival in CP was really better than 50 years ago. Aging with CP has received more attention, and has been discussed in different review articles (Liptak, 2008; Klingbeil *et al.*, 2004; Roebroek *et al.*, 2009; Rapp and Torres, 2000). CP was defined as a non-progressive disorder (Rosenbaum *et al.*, 2007), but secondary health-related problems are apparent with aging: increased incidence of bowel and bladder dysfunction, urinary tract infections, gastroesophageal reflux disease, oral motor and dental disorders, fractures, hip subluxation, progressive musculoskeletal deformity and dysfunction, deterioration of functional ambulation, pain and fatigue have been reported, which might result in reduced ability to fully engage in activities, restriction in participation and lower quality of life (Liptak, 2008; Klingbeil *et al.*, 2004; Roebroek *et al.*, 2009; Rapp and Torres, 2000).

## TREATMENT

### ***Treatment Options***

CP is one of the leading causes of disability in childhood. Currently no treatment options are available to cure the brain damage, and therefore the goal of treatment is to use appropriate combinations of interventions to improve patients' neuromuscular function, capabilities and participation in daily life (Rosenbaum, 2003; Krigger, 2006; Goldstein, 2004). The choices of treatment options depend on the patients' specific symptoms and age, and include global strategies, physiotherapy, external aids, oral or parenteral medication, and surgery (Goldstein, 2004; Rosenbaum, 2003; Sussman and Aiona, 2004; Koman *et al.*, 2004; Krigger, 2006; Steinbok, 2006; Tilton, 2009).

The major motor impairment seen in CP is spasticity which is found in 80 percent of cases (Stanley *et al.*, 2000b) and this leads to secondary abnormalities such as bony deformity, muscle and joint contractures (Koman *et al.*, 2004; Krigger, 2006). Therefore most of the management options aim to reduce spasticity in the early childhood to prevent secondary problems during lifespan. However, it has to be noted that there are some exceptions whereby the spasticity is "useful" and of functional benefit for the patient, and should then not be interrupted (Tilton, 2009).

### ***Spasticity Reduction***

The management of spasticity starts with in-depth evaluation with a patient's caregivers, physicians, allied health professionals and surgeons. As described by Tilton (2009) in a recently published review article, treatment should start with optimizing and maintaining joint range of motion by physiotherapy, stretching and orthotics. Thereafter the spasticity management team can decide to start with medication, neurosurgery, and/or consider orthopaedic surgery when the patient's gait is mature (Tilton, 2009). The choice of these treatment options depends on whether a reversible or permanent effect is desired, and if the spasticity is focal or general. Oral medication (e.g. baclofen, tizanidine, dantrolene), intrathecal baclofen (ITB) and botulinum toxin injections (botox) are reversible options, but botox is used for increased focal spasticity and oral medication and ITB for general spasticity. Orthopaedic surgery and neurosurgery both lead to permanent results: orthopaedic operations deal very effectively with the peripheral manifestations of spasticity, while

neurosurgery has the advantage of directly addressing the underlying neurological disorder (Ward, 2002). Of these, the neurosurgical procedure known as selective dorsal rhizotomy (SDR) has particular appeal as it interrupts the abnormal reflex circuit that maintains spasticity.

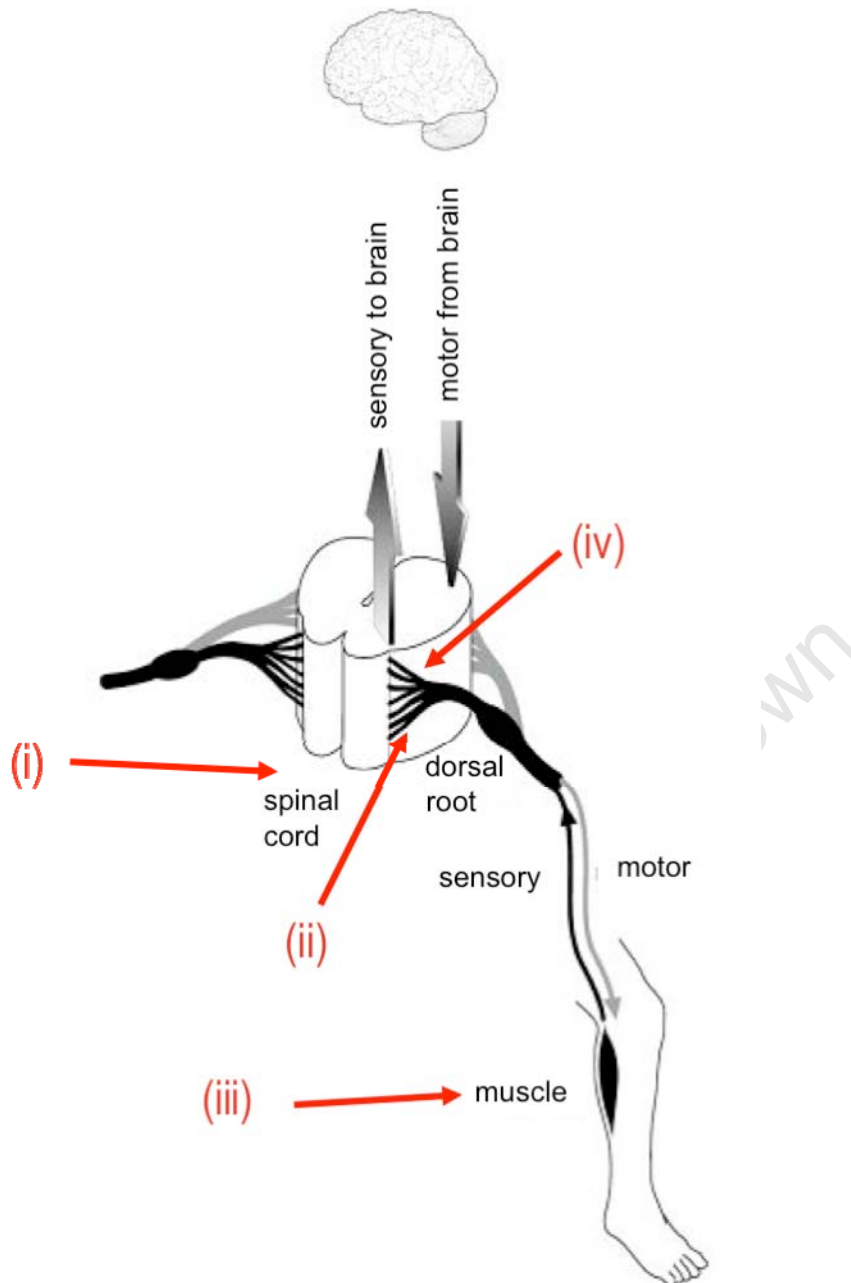
## **SELECTIVE DORSAL RHIZOTOMY**

### ***History***

Selective dorsal rhizotomy (SDR) is a neurosurgical procedure that reduces lower extremity spasticity by transecting a percentage of lumbar rootlets and disrupting the reflex arc at the spinal cord level. The history of SDR starts in the literature in 1898, when an English neurophysiologist called Sherrington first published an article about inducing spasticity in cats by mid-brain transaction (Sherrington, 1898). Thereafter, in 1908 the German neurosurgeon Otfried Foerster (1913) first performed SDR in humans. The technique he adopted was the division of the entire dorsal sensory roots from L2 to S1, excluding L4 to preserve knee extensor tone. This method fell into disuse because there was extensive sensory loss resulting from indiscriminate sectioning of the dorsal roots. In the 1960s in Montpellier in France, Gros (1967) revised the procedure by sectioning a fraction of the rootlets, while in the 1970s Fasano (1976; 1978) further refined the procedure by electrically stimulating the rootlets and measuring the electromyographic response prior to cutting the rootlets. In the 1980s in Cape Town, the neurosurgeon Warwick Peacock shifted the site of rhizotomy from the conus medullaris region to the cauda equina (Figure 1.1) for treating children with CP (Peacock and Eastman, 1981; Peacock and Arens, 1982). Having operated on more than 100 children, Peacock relocated from South Africa to Los Angeles in 1986 and, under his leadership, dorsal rhizotomy became accepted and used worldwide (Hesselgard *et al.*, 2007).

### ***Variation in Techniques***

Since the 1990s different adaptation of the 'Peacock' technique have been discussed. To restore spinal integrity laminotomies instead of laminectomies (Cochrane and Steinbok, 1992) and the optimal number of rootlets that should be sectioned (Kim *et al.*, 2002; McLaughlin *et al.*, 2002) were described. In addition, Tae



**Figure 1.1** 'Peacock' SDR technique: (i) laminectomy from L2 to S1 and opening of the dura; (ii) stimulate the posterior rootlets with a 50-Hz train of stimuli at the threshold intensity for muscular contraction; (iii) measure the muscle response with electromyography; and (iv) nerve rootlets associated with a normal response left intact, while those associated with an abnormal response divided (about 50-70% of L2-S1) (adapted from [www.uwhealth.org](http://www.uwhealth.org)).

Sun Park developed a technique where the number of laminectomy levels was reduced immediately caudal to the conus medullaris, which had to be localized by ultrasound (Park *et al.*, 1993; Park and Johnston, 2006). Steinbok questioned the electrophysiologic techniques as used during SDR (Steinbok and Kestle, 1996), and based on a 1 year follow-up study concluded that it is still advisable to use electrostimulation for guidance during the neurosurgical procedure, although there was not a significant difference in outcome with and without the use of this

stimulation (Steinbok *et al.*, 2009). Although different variations of the technique have been proposed, the 'Peacock' method (Peacock and Eastman, 1981; Peacock and Arens, 1982) is still the most popular technique and is performed in almost two-thirds of 36 different centres around the world (Hesselgard *et al.*, 2007).

### ***Patient Selection***

The main goal of SDR is to improve patients' function by releasing spasticity. In the 1980s Peacock *et al.* operated on patients classified as spastic (quadriplegia, triplegia and diplegia) and with dyskinesia (athetosis and dystonia). Based on their own follow-up studies they concluded that children with dyskinesia should not undergo SDR. The preference for patient selection is for children with spastic CP and predominantly leg involvement, with a certain level of intelligence and enough underlying muscle strength. The goal for the non-ambulatory patients was to ease daily handling for care givers (Peacock *et al.*, 1987; Arens *et al.*, 1989).

Currently the focus in our Department of Paediatric Neurosurgery is mainly on ambulant children, classified as spastic diplegia with or without minor involvement of unilateral upper limb and preferably aged between 3 and 8 years. In addition, patients are only selected if they have CP of congenital origin, good underlying muscle strength with trunk and motor control and absence of dyskinesia. One very important factor which should not be underestimated, is to ensure that the child receives intensive physiotherapy treatment before and after SDR. In addition, the motivation of the child and family/care-givers is crucial to success.

## **OUTCOMES OF SELECTIVE DORSAL RHIZOTOMY**

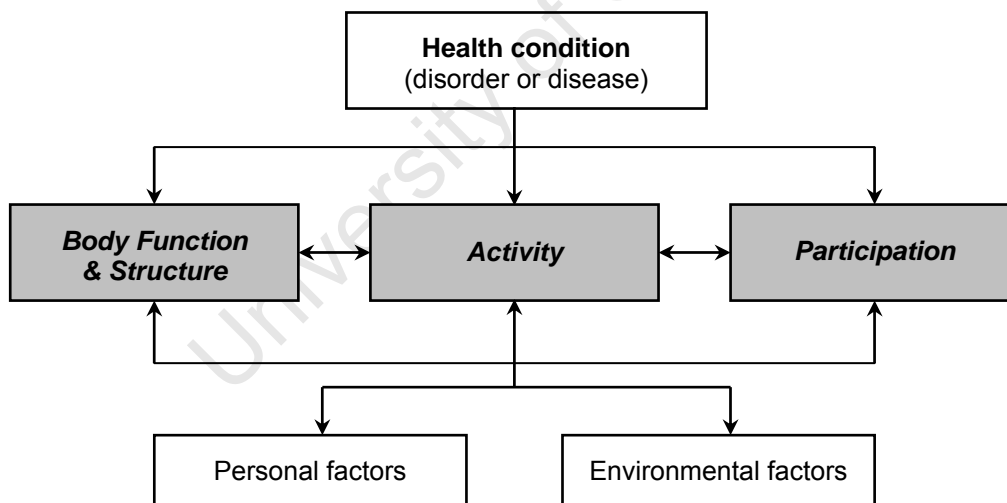
### ***International Classification of Functioning, Disability and Health (ICF) model***

To establish an understanding of the outcomes of SDR, outcome assessments should be identified and quantified across a broad spectrum. In 1980 the World Health Organisation (WHO) published the International Classification of Impairments, Disabilities and Handicaps (ICIDH), which was a classification of the "consequences of disease," developed to code a wide range of information about various aspects of health (WHO, 1980). In the early 1990s an international effort to revise and reshape

this classification system was initiated. After 9 years of study and input, the WHO published a new classification system, the International Classification of Functioning, Disability and Health, or ICF (WHO, 2001). The ICF focuses on the “components of health” rather than on the consequences of disease.

This ICF-model (Figure 1.2) provides a broad perspective within which to appreciate the spectrum of functioning and disability across the lifespan. It seeks to identify and classify abnormalities across two components: (i) *Body Function & Structure*; and (ii) *Activities and Participation*. These components are defined as follows:

- Body structures:* anatomical parts of the body such as organs, limbs and their components.
- Body functions:* physiological functions of body systems (including psychological functions).
- Activities:* execution of a task or action by an individual.
- Participation:* involvement in a life situation.



**Figure 1.2** The model based on the International Classification of Function (ICF) as published by the World Health Organization (WHO, 2001).

The ICF also recognized the importance of “contextual factors” that may act as obstacles or facilitators in shaping the level of functioning and disability, including Environmental and Personal factors. Environmental factors make up the physical, social and attitudinal environment in which people live and conduct their lives, while

personal factors are related to the person which have an impact on functioning (e.g. lifestyle, social background, education, life events, race/ethnicity).

Taking these two components and contextual factors into account, the ICF-model provides a biopsychosocial framework to identify and quantify clinical trials (Brockow *et al.*, 2004; Stucki *et al.*, 2002; Stucki, 2005) and has proven to be useful in the field of CP (Majnemer and Mazer, 2004; Rosenbaum and Stewart, 2004; Liptak, 2008).

### ***Body Function & Structure***

Steinbok (2001) published a review article based on 63 articles (published between 1966 and 1999) about the outcomes of SDR for patients with spastic CP. These outcomes were grouped in line with the model developed by the National Center for Medical Rehabilitation Research (NCMRR), which he revised in line with the ICF-model in 2007 (2007). In this same journal Farmer and Sabbagh (2007) also published a review regarding SDR. Both articles provided evidence that SDR leads to improvements in the ICF-model *Body Function & Structure* component during the first few years after surgery. Taking into account the articles published in the last 2 years (Chan *et al.*, 2008; Cole *et al.*, 2007; Nordmark *et al.*, 2008; Trost *et al.*, 2008), it can be concluded that in the first years after SDR ( $\leq 10$  years), children showed decreased muscle tone, increased passive and active joint range of motion, and improved motor control, which resulted in an improved gait pattern. However, the influence of SDR on muscle strength is debatable and has been reported to deteriorate, remain unchanged, or improve during the first years after surgery (Steinbok, 2001; Farmer and Sabbagh, 2007). Since SDR prevents secondary abnormalities, some studies have shown that it can also reduce the number of necessary orthopaedic surgeries (Hagglund *et al.*, 2005; O'Brien and Park, 2006), while others have raised their concerns regarding a continuous need for orthopaedic surgeries (Aiona and Sussman, 2004).

SDR was also compared to other interventions, which provided positive statements in favour of this neurosurgical procedure. McLaughlin *et al.* (2002) published a meta-analysis of 3 randomized controlled trials wherein the outcomes of SDR with physiotherapy (SDR+PT) was compared to only physiotherapy (PT) in children with spastic diplegia 9 or 12 months after intervention. It was unanimously concluded that

muscle tone (Ashworth scale) in the SDR+PT was significant lower than in the PT group. Engsborg *et al.* (2006) also evaluated outcomes of children with spastic diplegia who underwent SDR plus physiotherapy compared to those receiving only physiotherapy. They showed decreased spasticity in the SDR+PT group 20 months after SDR, while the PT group did not show any changes. Muscle strength did improve in both groups, but the gait kinematics and walking speed showed greater improvement in the SDR+PT than the PT group. Kan *et al.* (2008) compared, based on a 1 year follow-up, the reversible and irreversible neurosurgical options intrathecal baclofen in patients with severe preoperative spasticity (GMFCS  $\geq 3$ ). They concluded that greater improvements were seen for muscle tone (Modified Ashworth scale) and passive joint range of motion in the SDR group, and fewer additional orthopaedic surgeries were necessary. The difference in outcomes between SDR and orthopaedic surgery was assessed by Abel *et al.* (1998). They completed a 1 year follow-up study in children with spastic diplegia, wherein temporal-distance parameters (as a result of 3-dimensional gait analyses) were compared. Based on change in cadence, they concluded that muscle tendon lengthening showed a more robust positive change in walking pattern than SDR. However, Thomas *et al.* (2004) found no significant difference between the gait kinematic data 2 years after SDR between patients who received SDR or orthopaedic surgery. In addition, they found similar improvements in muscle tone (Ashworth scale), passive range of motion and oxygen consumption for both surgical procedures. The Portland research group (Buckon *et al.*, 2004) also published their findings on children's functioning 2 years after they underwent SDR or orthopaedic surgery. They showed, based on the Gross Motor Performance Measure (GMPM), that both interventions improved alignment, weight shift, and stability, although the SDR group demonstrated more prevalent qualitative changes in movement.

### ***Activity and Participation***

With regards to the ICF-model, Steinbok (2001; 2007) concluded in his review article that strong, although inconclusive, evidence was found for improvements in outcomes of *Activity*, whereas only moderate evidence was found for a reduction in the level of *Participation*. Farmer and Sabbagh (2007) also reported the benefits of SDR in this ICF component based on Gross Motor Function Measure (GMFM), Peabody Developmental Motor Scale (PDMS), Pediatric Evaluation of Disability

Inventory (PEDI) and Functional Independence Measure for Children (WeeFIM). In addition, they concluded that young children who were (potentially) ambulant had a higher chance of achieving functional benefit from selective dorsal rhizotomy than older and more involved children. After the publications of these two review articles in 2007, Chan *et al.* (2008) and Nordmark *et al.* (2008) confirmed these positive outcomes of GMFM and PEDI after SDR. Nordmark *et al.* showed that the largest changes were seen in children classified at GMFCS levels I and II 5 years after surgery. Cole *et al.* (2007) reported functional improvement 5 years after SDR based on changes in GMFCS levels, although the GMFCS levels have been developed as a classification system and *not* as an outcome measure (Palisano *et al.*, 1997).

In comparison to other interventions, SDR leads to fewer limitations in activities and restrictions in participation. The meta-analyses, wherein GMFM levels for SDR+PT and PT groups were compared, showed small but statistically improved results in the SDR+PT group (McLaughlin *et al.*, 2002). Engsborg *et al.* (2006) also found improvements in GMFM in both groups, while the increase in Gross Motor Ability Estimate (GMAE) was only significantly higher in the SDR+PT compared to the PT group. The study wherein outcomes of SDR and ITB were compared, showed a significantly improved change in GMFCS levels in the SDR group (Kan *et al.*, 2008). Based on the GMFM Abel *et al.* (1998) found that 1 year postoperative patients who underwent SDR had more global positive changes in functional mobility and health related quality of life, than children who only received muscle tendon lengthening only. Similarly, Buckon *et al.* (2004) showed greater improvement in the SDR group than children who received orthopaedic surgery 2 years after surgery, although orthopaedic surgery might have had a more rapid improvement in gross motor skills (GMFM). In addition, regarding self-care, mobility, social skills and ability to achieve independence (PEDI), the SDR group developed more rapidly and with greater frequency than the orthopaedic group.

### **Complications**

Despite the fact that SDR almost invariably succeeds in its aim of reducing spasticity, various complications may occur as a result of the surgical approach. The most notable of these is spinal abnormality brought about by the laminectomy performed in order to allow access to the dorsal roots (Aiona and Sussman, 2004; Steinbok,

2007). Examples are an increased incidence of scoliosis, thoracic hyperkyphosis, lumbar hyperlordosis, spondylolysis and spondylolisthesis (Peter *et al.*, 1990; Johnson *et al.*, 2004; Steinbok *et al.*, 2005; Turi and Kalen, 2000; Spiegel *et al.*, 2004; Golan *et al.*, 2007; Peter *et al.*, 1993). In relation to these spinal deformities, various short-term follow-up studies have reported problems of back pain (Steinbok and Schrag, 1998; Spiegel *et al.*, 2004; Johnson *et al.*, 2004; Turi and Kalen, 2000). Another concern that has been identified is progressive subluxation of the hips, although this does not normally deteriorate as a result of SDR. Other complications, some of which are directly related to the procedure itself, include bronchospasm, bladder and bowel dysfunction, postoperative pain and sensory alteration, although these can be avoided by administering suitable medication (Steinbok, 2007; Farmer and Sabbagh, 2007).

## **OUTLINE OF THE THESIS**

### ***Research Question and Aims***

Many studies have shown that there is indeed a reduction in spasticity and improvement in function as a result of SDR, although most of these studies were based on assessments performed during the first years after surgery. Therefore, the question remains: what will happen when these patients with CP become adults?

Given the large number of patients with CP who have undergone SDR in the past three decades, there is an important clinical need to evaluate the long-term effects of this procedure (van Schie *et al.*, 2005; Farmer and Sabbagh, 2007). Since we in Cape Town reintroduced this procedure and have a long track record of research in this field (Berman, 1989; Berman *et al.*, 1990; Peacock and Eastman, 1981; Peacock and Arens, 1982; Peacock *et al.*, 1987; Peter *et al.*, 1990; Peter *et al.*, 1993; Peter and Arens, 1993; Peter and Arens, 1994; Subramanian *et al.*, 1998; Vaughan *et al.*, 1988a; Vaughan *et al.*, 1988b; Vaughan *et al.*, 1989; Vaughan *et al.*, 1991; Vaughan *et al.*, 1998), we have a responsibility to the clinical community, patients (with special focus on patients who are classified as spastic diplegia, with or without minor involvement of unilateral upper limb) and parents to answer some of these important questions.

Therefore the aim of this doctoral thesis is to investigate the long-term sequelae of SDR in patients with spastic diplegia seen in the context of the broad spectrum of the ICF-model *Body Function & Structure, Activities and Participation*.

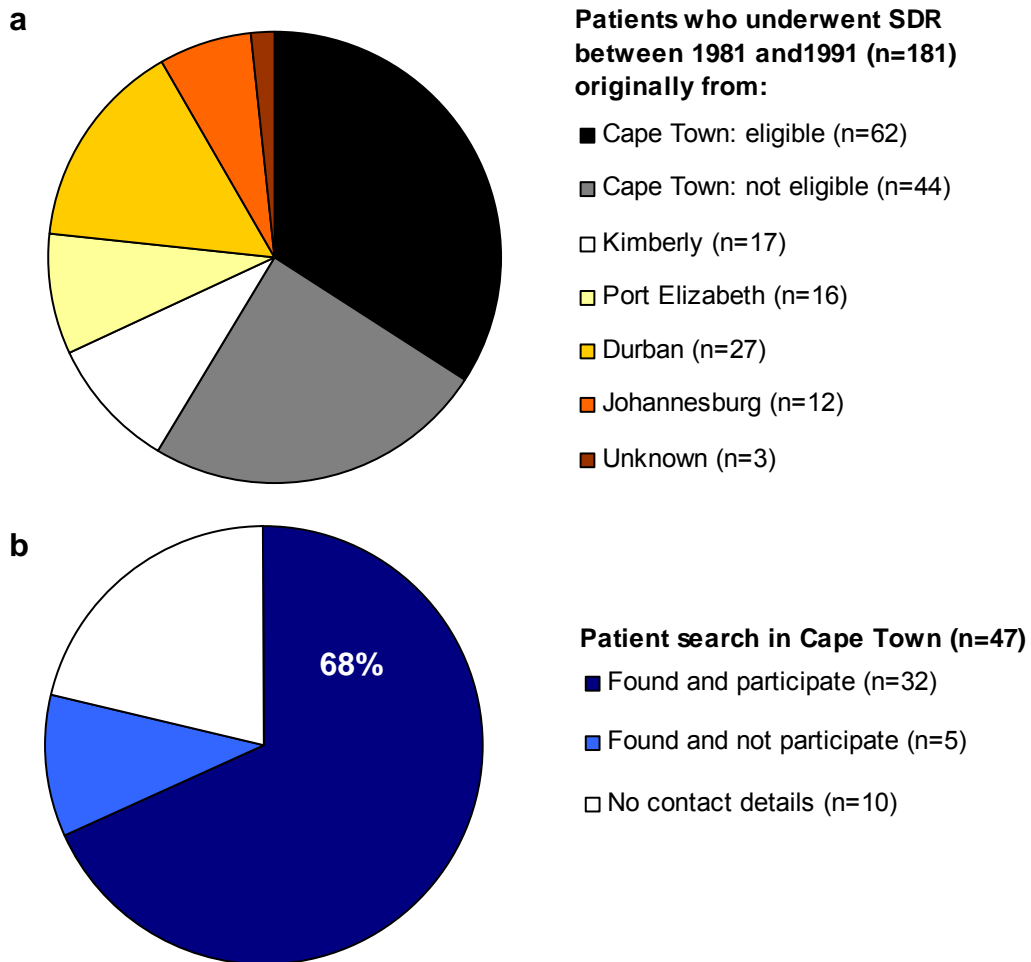
### Projects

This thesis is based on two main research projects. Project I included 20 year follow-up studies after SDR in patients with spastic diplegia. These studies were in line with our former short-term follow-up studies based on 14 children with spastic diplegia who received SDR in 1985 (Berman, 1989; Vaughan *et al.*, 1988b; Vaughan *et al.*, 1991; Subramanian *et al.*, 1998). Contact details were no longer held by Red Cross Children's Hospital where they were all originally treated, and a central CP register does not exist in South Africa.

Therefore an extended search was necessary. In 2005, 100% of the study cohort was tracked down and a road trip around South Africa (Cape Town to Johannesburg to Durban to Port Elizabeth and back to Cape Town) was completed (Figure 1.3), during which data collection was performed.



**Figure 1.3** Visit of patients participated in project I located in South Africa: [1]: Johannesburg, n=2; [2]: Pietermaritzburg, n=1; [3]: Durban, n=2; [4]: Port Elizabeth, n=1; [5]: Calitzdorp, n=1; [6]: Paarl; n=1; and [7]: Cape Town, n=7 (adapted from www.maps.com).



**Figure 1.4** Distribution of places where patients lived when they underwent SDR in South Africa (majority underwent the operation in Cape Town) between 1981 and 1991 (a) and which portion of the eligible patients living in or close to Cape Town did participate in project II (b).

Project II is based on patients who were operated on between 1981 and 1991 and participated in earlier studies with a focus on spinal deformity (Peter *et al.*, 1993; Peter *et al.*, 1990). In these 10 years 181 children underwent SDR in South Africa, with the majority undergoing their operation in Cape Town. As shown in Figure 1.4a, 106 patients lived in Cape Town at the time the operation was performed, with 62 eligible for this project. Again no contact details were available, except for the 8 adults who also participated in the first project and lived in Cape Town or within a radius of 100 kilometres at the time of data collection. With help from clinicians, patients, patients' families, teachers and physiotherapists at schools and an internet search, we completed an intensive search. We found that 15 patients were not living in or close to Cape Town at the time of data collection, and so 47 eligible patients remained. As presented in Figure 1.4b, we were able to track down 37 of these adults, with 5 of them not able to participate in this project (not based on health

reasons). Finally, 32 patients with spastic diplegia (with or without mild involvement of the unilateral limb) were included in this second long-term follow-up project.

### **Studies**

The 20 year follow-up studies (project I) are described in Chapters 2, 3 and 4, while the outcomes of project 2 are reported in Chapters 5 and 6.

Chapter 2 addresses the long-term experience with SDR of our research group in Cape Town. Outcomes of our former published studies regarding SDR are summarized and general experiences from the 20 year follow-up study described. This overview was published in a special edition on selective dorsal rhizotomy in the journal *Child's Nervous System* in 2007.

In line with the 1, 3, and 10 year follow-up gait studies, we conducted a 20 year follow-up study with 2-dimensional gait analysis as the primary outcome measure. This study is presented in Chapter 3 and was published in the *Journal of Neurosurgery: Pediatrics* in 2008.

Chapter 4 includes, in line with our 1 year follow-up gait study, a 20 year follow-up study with a focus on functional outcome measures. This chapter was published in *Archives of Physical Medicine and Rehabilitation* in 2009.

As a result of project II long-term outcomes regarding spinal abnormalities are described in Chapter 5. Besides the results of X-ray and Magnetic Resonance Imaging (MRI) scans, some clinical assessments are described for these patients who underwent SDR between 17 and 26 years ago. This article was accepted by *Child's Nervous System* in August and will be published in December 2009.

Chapter 6 is focused on patients' activity and participation in the community 17 to 26 years after SDR. This study is on functional mobility (Functional Mobility Score: FMS) and accomplishment and satisfaction levels over a wide range of life habits (LIFE-H 3.1 questionnaire), and has been submitted to *Disability and Rehabilitation*.

The thesis concludes with a general evaluation of the reported findings and discussion in the light of the ICF-model (Chapter 7). In addition some future directions for research are outlined.

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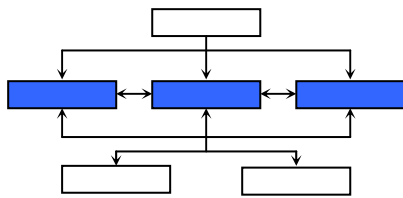
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# Chapter 2



## Selective dorsal rhizotomy: long-term experience from Cape Town

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*Child's Nervous System*, 23(9): 1003-1006, 2007

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## ABSTRACT

**Introduction:** Given the large number of cerebral palsy patients who have undergone selective dorsal rhizotomy in the past two decades, it is clearly imperative that the clinical community be provided with objective and compelling evidence of the long-term sequelae of the procedure. **Materials and methods:** In the early 1980's, Peacock in Cape Town shifted the site of the rhizotomy from the conus medullaris to the cauda equina, and in the past 25 years more than 200 children have been operated on. We have studied the incidence of spinal deformities after multiple level laminectomy and recorded a 20% incidence of isthmic spondylolysis or grade I spondylolisthesis. We have also conducted a long-term prospective gait analysis study on a cohort of 14 ambulatory patients who were operated on in 1985. **Results:** Ten years after surgery, our patients had increased ranges of motion that were within normal limits. Step length was significantly improved, although cadence was unchanged postoperatively and was significantly less than normal age-matched control subject. **Discussion:** We have recently tracked down all 14 patients from the original cohort and are currently completing a 20 year prospective follow-up analysis of their neuromuscular function and gait. Our preliminary data suggest that selective dorsal rhizotomy is not only an effective method for alleviating spasticity, but it also leads to long-term functional benefits.

**Keywords:** Children • Cerebral palsy • Dorsal rhizotomy • Spinal deformity • Gait analysis

## INTRODUCTION

With increased survival of low-birth-weight infants and increased longevity of the adult population, the number of adults with cerebral palsy (CP) is increasing. Klingbeil *et al.* (2004) reviewed the problems of aging in several groups of individuals with disabilities, including CP. Although CP is normally described as a non-progressive disorder, cumulative evidence over the past two decades supports the patients' perspective of functional deterioration associated with aging. There are extra concerns regarding selective dorsal rhizotomy (SDR) and its long-term effects on the potential production of muscle weakness, the possible development of spinal deformities, and the continuing need for orthopaedic intervention (Aiona and Sussman, 2004). Given the large number of CP patients who have undergone SDR in the past two decades, particularly in the United States, it is clearly imperative that the clinical community be provided with objective and compelling evidence of the long-term sequelae of the procedure. With our long record in Cape Town we are well-placed to provide that evidence.

## MATERIAL AND METHODS

Although dorsal rhizotomy was described almost 100 years ago by the German surgeon Otfried Foerster (1913), it fell into disuse for over 50 years because of the adverse consequences of deafferentation. In the 1960's the procedure was revised by sectioning a fraction of the rootlets (Gros *et al.*, 1967) and then further refined into *selective* dorsal rhizotomy by electrically stimulating the rootlets and measuring the electromyographic response prior to cutting the rootlets (Fasano *et al.*, 1976). In Cape Town, Peacock and Eastman (1981) shifted the site of rhizotomy from the conus medullaris region to the cauda equina and reintroduced the technique in the early 1980's for treating children with cerebral palsy.

Having performed more than 100 such operations, Peacock relocated to Los Angeles in 1986, and his contribution led to the use of selective dorsal rhizotomy becoming widespread in the USA (Abbott *et al.*, 1989; Oppenheim, 1990; Park and Owen, 1992; Steinbok, 2001). Since Peacock's departure in 1986, SDR has continued in

Cape Town, first conducted by Jonathan Peter and more recently by Graham Fieggen. In the past 25 years more than 200 children have undergone SDR in Cape Town.

Peter *et al.* (1990) studied the incidence of spinal deformities in children after multiple level laminectomy for SDR and concluded that spondylolysis was the only abnormality that appeared to be more common in patients who underwent the procedure compared with those who did not have SDR. They felt that it was essential to maintain an ongoing follow-up of the spines of post-rhizotomy patients, both to monitor the children identified with spondylosis and spondylolisthesis and also to determine the true incidence of this condition in the whole SDR group. They also sought to determine whether these conditions increased with time. In their study which included 163 children, the patients had five-level lumbosacral laminectomies, and 99 were X-rayed postoperatively. Of these, 20% developed incidental isthmic spondylolysis or grade I spondylolisthesis, leading Peter *et al.* (1993) to postulate that the laminectomy, associated lordosis and increased mobility after SDR may all be causative factors.

In an effort to reduce the problems of lumbar instability (Peter *et al.*, 1990), some surgeons have adopted a more limited laminectomy at the level of the conus as advocated by Park (2000). There is also some controversy as to the *number* of rootlets that should be sectioned, where less than 25% can lead to limited functional gains (McLaughlin *et al.*, 1994) but Kim *et al.* (2001) has also suggested that more than 50% can also be problematic.

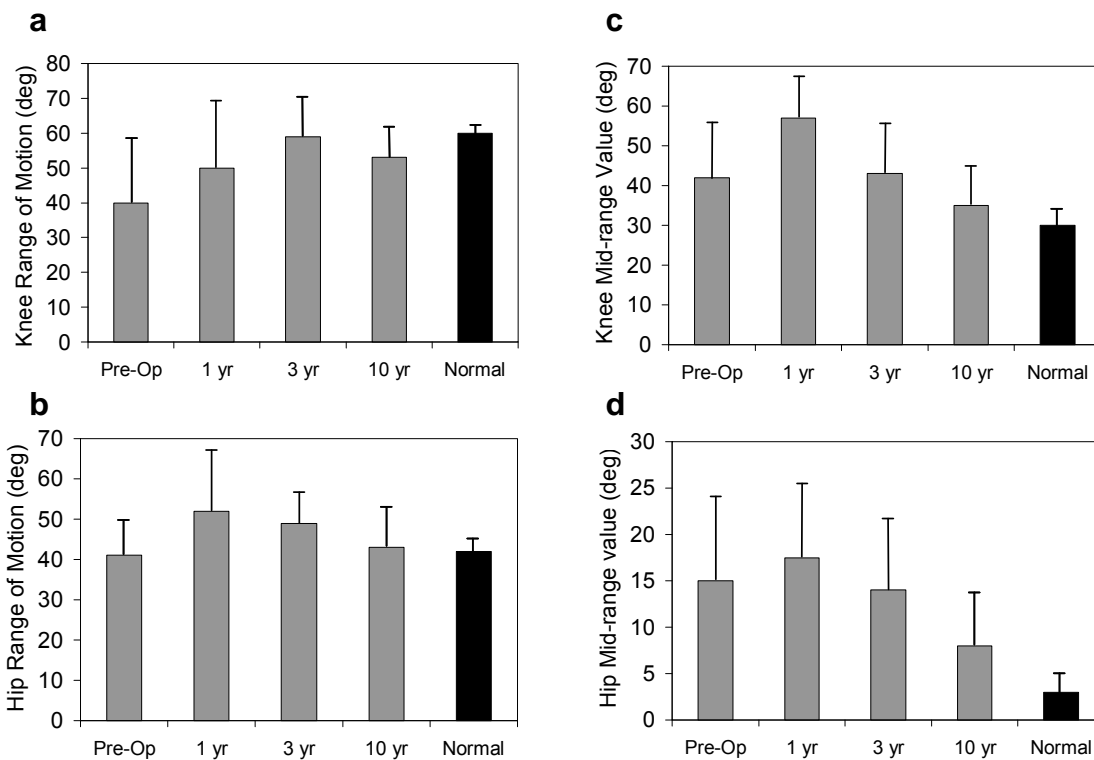
In 1985 Berman, Vaughan and Peacock introduced functional assessments that were conducted preoperatively and one year post-surgery (Berman *et al.*, 1990). Since the Gross Motor Function Measure (GMFM) scoring system had not yet been developed (Russell *et al.*, 2004), they proposed their own rating scales (Berman, 1989). These scales were designed to test muscle tone, voluntary movement, joint range of motion, and functional movement in certain developmental positions. The results of this study indicated positive gains after SDR, although this was dependent on the patients' abilities before surgery (Berman, 1989; Berman *et al.*, 1990). One of the greatest gains made was in the patients' ability to assume and maintain certain postures,

which, in turn, allowed for improved participation in activities. It was also obvious that once spasticity had been removed, the CP patients had greater freedom of movement (Berman *et al.*, 1990).

Twenty years ago the Cape Town team, led by Vaughan, was the first group to study the effects of SDR with gait analysis. In their original study, 14 ambulatory patients were involved in a gait analysis follow-up study one year after surgery (Vaughan *et al.*, 1988; Vaughan *et al.*, 1989). The study was then repeated on a cohort of 11 patients two years later as a three year follow-up study (Vaughan *et al.*, 1991) and also 10 years postoperatively (Subramanian *et al.*, 1998; Vaughan *et al.*, 1998). The parameters they used to substantiate their findings were joint kinematics (hip and knee ranges of motion and hip and knee midrange values) and temporal-distance parameters (step length, cadence and velocity). To accommodate musculoskeletal growth, the temporal-distance parameters were normalized to yield dimensionless numbers according to the method of Hof (1996).

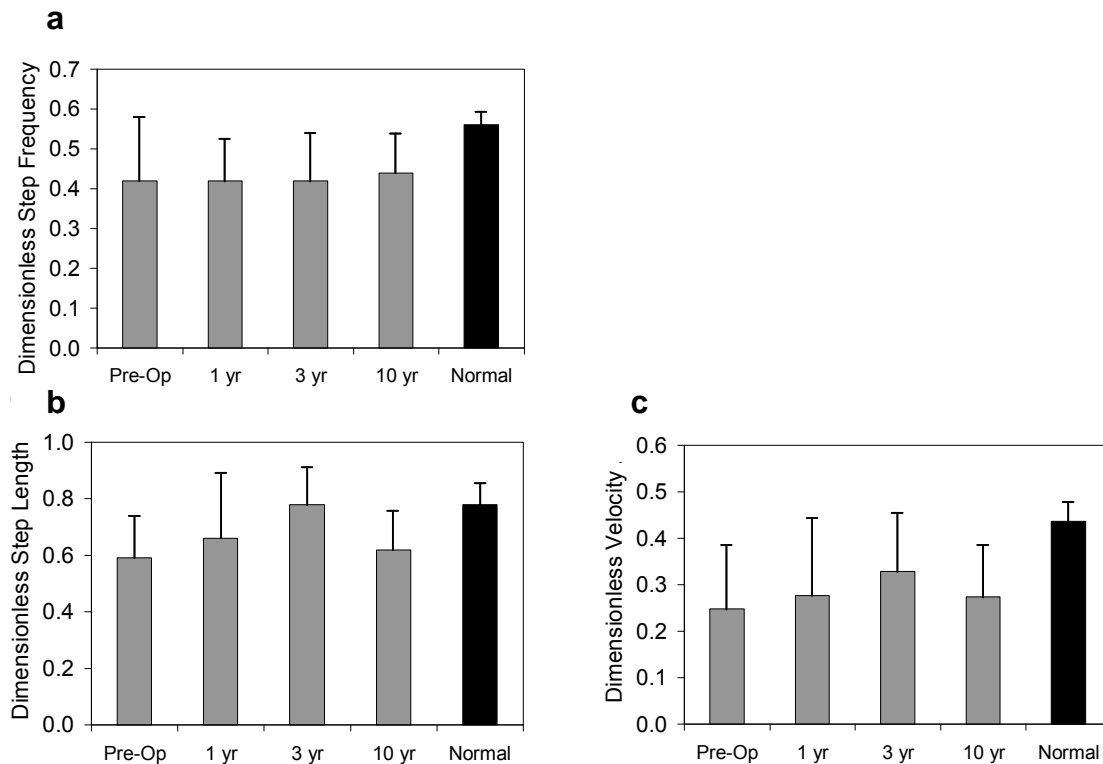
## RESULTS

Figure 2.1 presents the ranges of motion and midrange (*i.e.*, the angle of the knee when the leg is at its mid-point in stance phase) values for the knee and hip joints. The range of motion at the knee and hip increased as a result of surgery, and by 10 years there was no statistical difference from normal ranges. Concomitant with the increased ranges of motion, there was also the deleterious increase in midrange values immediately after surgery. This was of some concern because the midrange values represent a measure of strength and control exhibited by a patient during locomotion. The greater the midrange value, the more flexed the posture of the individual, and this resulted in an exaggerated crouch gait. However, these midrange values decreased over time, so that by 10 years postoperatively there was no variance from normal values. Thus, 10 years after surgery, patients not only had increased ranges of motion that were within normal limits, but they also used that functional movement at approximately a normal midrange point (Subramanian *et al.*, 1998).



**Figure 2.1** Sagittal plane joint kinematics: (a) Knee range of motion; (b) Hip range of motion; (c) Knee midrange value; and (d) Hip midrange value, for 11 subjects before selective dorsal rhizotomy, and at 1, 3 and 10 years after surgery compared with normal controls (Subramanian *et al.*, 1998; Vaughan *et al.*, 1998).

The mean data for the normalized temporal-distance parameters of the 10 year follow-up study are presented in Figure 2.2 (Subramanian *et al.*, 1998). The dimensionless cadence (step frequency) was unchanged postoperatively, and was always significantly less than normal (Figure 2.2a). Since cadence is likely to be centrally mediated, and SDR attacks the problem at the level of the spinal cord, it is not surprising that the procedure has a limited influence on this parameter (Vaughan *et al.*, 1998). Figure 2.2 also shows the benefits of SDR in significantly improved step length as a natural consequence of the release of spasticity and the increase in knee range of motion and hip range of motion. Ten years after surgery there is a small deterioration, although this may have resulted from the fact that five of the 11 subjects were videotaped in a confined room that was not conducive to attaining a standard gait. Velocity is the product of cadence and step length, and shows a trend that is comparable to step length after surgery, because cadence does not change postoperatively (Subramanian *et al.*, 1998).



**Figure 2.2** Dimensionless temporal-distance parameters: (a) cadence or step frequency; (b) step length; and (c) velocity, for 11 subjects before selective dorsal rhizotomy, and at 1, 3 and 10 years after surgery compared with normal controls (Subramanian *et al.*, 1998; Vaughan *et al.*, 1998). Note that the parameters have all been normalized according to Hof (1996).

## DISCUSSION

In line with our original functional assessment and gait analysis studies performed in 1985, our Cape Town research group has recently conducted a 20 year prospective follow-up study. We have repeated the tests used in our previous functional (Berman *et al.*, 1990; Berman, 1989) and gait (Vaughan *et al.*, 1988; Vaughan *et al.*, 1991; Subramanian *et al.*, 1998) studies and the data are currently being analyzed. We were able to track down the original cohort of 14 spastic diplegic cerebral palsy patients who underwent SDR by Peacock. The mean age of these 14 patients in 2005 was 27.9 years (range 22.1 - 33.0) with a gender distribution of 6 females and 8 males.

Besides participating in the functional assessment and gait studies, we requested the 14 patients to complete a questionnaire regarding details about their past and

present life, health status and their outlook on SDR. Twelve of the cohort had a positive outlook on SDR, while two had mixed feelings. Both of them were suffering from muscle weakness postoperatively, which resulted in one of them becoming wheelchair-bound. Although all patients received physical therapy at school preoperatively, more than half of the group needed tendon lengthening before SDR. After rhizotomy another 4 underwent this orthopaedic procedure, while half of them required osteotomy. More than half the cohort was suffering from some back pain, while 2 of the patients walked with a crutch and cane. In answer to the question "Do you need help with activities of daily living?" all 14 patients answered "No." Only 2 of them were not employed or studying at college. We established that besides medical intervention, it is very important that the patients have a good understanding of their specific motor problems, and that they are supported by family and friends who help to motivate them.

From this series of follow-up studies in Cape Town it has been shown that SDR is an effective method for alleviating spasticity, and the team has provided evidence to show that functional benefits, as measured by improved gait, can also be obtained (Subramanian *et al.*, 1998). Our Cape Town research group would appear to be the only authors to have published long-term (greater than 5 years) gait analysis studies on the efficacy of SDR.

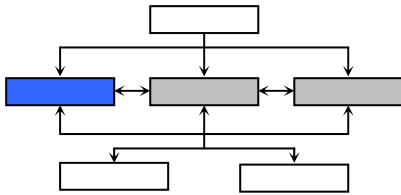
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# Chapter 3



A prospective gait analysis study in patients with  
diplegic cerebral palsy 20 years after  
selective dorsal rhizotomy

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## ABSTRACT

**Object:** Selective dorsal rhizotomy (SDR) has been widely performed for the reduction of spasticity in patients with cerebral palsy during the past 2 decades. The objective of this study was to determine whether the surgery has yielded long-term functional benefits for these patients. **Methods:** We present here results from a prospective 20 year follow-up study of locomotor function in 13 patients who underwent an SDR in 1985. For comparison, we also present gait data for 48 age-matched healthy controls (12 at each of 4 time points). Patients were studied preoperatively, and then at 1, 3, 10 and 20 years after surgery. Study participants were recorded in the sagittal plane while walking using a digital video camera, and 6 standard gait parameters were measured. **Results:** In this group of patients 20 years after surgery, knee range of motion (ROM) was on average 12° greater than preoperative values ( $p < 0.001$ ). Hip ROM before surgery was no different from that in the healthy control group. This parameter increased markedly immediately after surgery ( $p < 0.001$ ) but had returned to normal after 20 years. The knee and hip midrange values — a measure of the degree of “collapse” due to muscle weakness after surgery — had returned to preoperative levels after 20 years, although they were respectively 11 and 8° greater than those in healthy controls. Both temporal-distance parameters (dimensionless cadence and dimensionless step length) were significantly greater at 20 years than preoperative values (cadence,  $p = 0.003$ ; step length,  $p = 0.02$ ), leading to improved walking speed. **Conclusions:** Twenty years after undergoing SDR, our patients showed improved locomotor function compared with their preoperative status.

**Keywords:** Cerebral palsy • Gait analysis • Long-term follow-up • Range of motion • Selective dorsal rhizotomy

## INTRODUCTION

The prevalence of cerebral palsy (CP) has risen from 1.5 per 1,000 live births in the 1960s to the current rate of 2.5 per 1,000 in developed countries (Odding *et al.*, 2006). In addition, there is increased longevity in the adult CP population (Kriger, 2006). Although CP is a complex neurological condition resulting from a *non-progressive* lesion in the developing brain (Bax *et al.*, 2005), adults with CP experience secondary complications and accelerated impairments (Klingbeil *et al.*, 2004). Because 80% of CP cases have spasticity as the primary abnormality (Stanley *et al.*, 2000), which can lead to secondary abnormalities such as muscle contractures (Gage, 2004), it is important to use insight when treating this condition. Treatment strategies for CP spasticity have included physical therapy, orthotic devices, oral pharmacological intervention, parenteral medication and surgery (Goldstein, 2004; Goldstein, 2004; Koman *et al.*, 2004; Steinbok, 2007; Sussman and Aiona, 2004). Only the intrathecal baclofen pump and selective dorsal rhizotomy (SDR) interfere directly with the primary abnormality of spasticity in CP by attempting to restore the balance of spinal circuits (Gage, 2004; Steinbok, 2001).

Selective dorsal rhizotomy was first performed by Foerster (1913) in the early 1900's and modified in the 1960's by Gros (1967). In the 1970's Fasano *et al.* (1976) further refined the procedure by cutting the posterior rootlets on the basis of intraoperative functional electrical stimulation of leg muscles. Peacock (Peacock and Eastman, 1981; Peacock and Arens, 1982) refined this method in the 1980's by exposing the cauda equina to identify dorsal rootlets at their exit foramina. Despite some controversy regarding its efficacy (Landau and Hunt, 1992), SDR has gained widespread acceptance over the past 2 decades (Aiona and Sussman, 2004; Gage, 2004; Park and Owen, 1992; Stansfield *et al.*, 2003; Steinbok, 2001; Steinbok, 2006). In a recent review, Steinbok (2001) concluded that there was strong evidence of decreased lower limb spasticity, increased lower limb range of motion (ROM) (without loss of muscle strength) and a moderate degree of improvement in activities of daily living after SDR for spastic CP. Although a large numbers of studies support the efficacy of SDR in reducing spasticity and improving function, most of these are based on assessments performed within 1 to 5 years after surgery. Our own research, based on a prospective study of Dr. Peacock's patients at 1, 3 and 10

years after surgery, has provided evidence to show that lasting functional benefits (as measured by improved gait) can be obtained with SDR patients with CP become adolescents and young adults (Berman *et al.*, 1990; Langerak *et al.*, 2007; Subramanian *et al.*, 1998; Vaughan *et al.*, 1991; Vaughan *et al.*, 1988; Vaughan *et al.*, 1989).

Given the large number of patients with CP who have undergone SDR in the past 2 decades, it is clearly imperative that the clinical community is given objective and compelling evidence of the long-term sequelae of the SDR procedure when these patients become adults. In this paper, we present results from a prospective 20 year follow-up study of 13 patients who received SDR surgery in 1985, with a particular focus on their gait data.

### **CLINICAL MATERIALS AND METHODS**

This study was approved by the Human Ethics Committee at the University of Cape Town, and each participant signed an informed consent form. In addition, the patients with CP completed a questionnaire that recorded other interventions, amount of physical therapy, problems with daily activities, and their personal feelings about the outcome of the SDR procedure.

Twenty-nine children with CP underwent SDR in 1985. Selection for surgery was based on the following criteria: (1) exhibition of spasticity; (2) absence of athetosis; (3) absence of significant underlying muscle weakness; and (4) access to postoperative therapy. Fourteen of the 29 children were able to walk (4 household ambulators and 10 independent ambulators) and participated in the first study comparing preoperative and 1 year postoperative gait analysis data (Berman *et al.*, 1990; Vaughan *et al.*, 1988; Vaughan *et al.*, 1989). Eleven of these 14 patients were available and participated in the 3 and 10 year follow-up studies (Langerak *et al.*, 2007; Subramanian *et al.*, 1998; Vaughan *et al.*, 1991). We located all 14 original patients in early 2005, and each was willing to participate in this reassessment. One of the 14 patients had severe muscle weakness and was not able to walk independently; for this reason, she was excluded from the study. Two patients were

able to walk using a device, 1 using a crutch and the other using a cane (Langerak *et al.*, 2007). The mean age of the 13 patients who participated in the 20 year follow-up study during 2005 was 27.3 years (range 22 to 34 years). For purposes of comparison, gait analysis was also performed on a group of 12 age-matched healthy controls who had a mean age of 27.8 years (range 22 to 34 years). Table 3.1 provides an overview of the groups which participated in the different studies.

### ***Selective Dorsal Rhizotomy***

All children underwent operations performed by Dr Peacock at the Red Cross Children's Hospital in Cape Town, South Africa. Each patient received endotracheal general anesthesia (without the use of long-acting neuromuscular blocking agents) and was then placed in a prone position with bolsters under the chest and pelvis. Limited laminectomies were performed from L2 to S1, exposing the cauda equina, with preservation of the posterior facet joints. The dorsal roots were separated from the anterior roots. For the electromyographic recordings, electrodes were placed in major muscle groups of the lower extremities, corresponding to the nerve rootlets tested. A 50 Hz train of stimuli was applied at the threshold intensity for muscular contraction and those dorsal nerve rootlets associated with a normal response were left intact, whereas those associated with an abnormal response were divided.

### ***Gait Evaluation***

Our 2D kinematic gait analysis system consisted of lightweight retroreflective markers, a linear calibration standard and a portable digital video camera (DCR-TRV80E, Sony). Although it would have been preferable to study all patients in the same laboratory, the cost of transportation precluded this option. Two researchers (NGL, RPL) drove by car with all the equipment throughout South Africa to visit 8 of the 13 CP patients in their home towns. The gait data were captured in a dimmed room with a walkway of at least 11 meters, similar to our laboratory in Cape Town. Three markers were placed at the hip (greater trochanter of the femur), knee (lateral femoral epicondyle) and ankle (lateral malleolus of the distal fibula) of each individual. These markers reflect the light transmitted by an infra-red light source on top of the camera. Separate analyses of the right and left lower limbs were performed in the sagittal plane. The patient was required to walk barefoot, without any orthoses, at his or her normal pace in a plane perpendicular to the camera's optical axis. The

data obtained from the camera were then analyzed digitally using customized software for gait analysis. Through suitable calibration and scaling, the x and y coordinates of the joints were obtained in real units (meters) as a function of time (sample rate of 25 Hz). From these data it was possible to calculate all relevant gait parameters.

### **Outcome Measures**

As in our previous pre- and postoperative studies, the gait parameters included angular kinematic data and temporal-distance parameters. The angle at the knee joint was defined as the angle between the thigh and the calf, whereas the hip angle was defined as the angle between the thigh and the vertical axis. Hip and knee midrange values were defined as the point half-way between the extremes of extension and flexion (Subramanian *et al.*, 1998; Vaughan *et al.*, 1991). The temporal-distance parameters included cadence (or step frequency, the number of steps per second) and step length (the distance in meters between 2 consecutive heel strikes; [left-to- right or right-to-left]).

When performing a longitudinal study on children it is important to account for the way that study participants grow and mature. Angular kinematic data do not change after a child reaches the age of 3 years and so these parameters are comparable in all follow-up studies. In contrast, temporal-distance parameters change as a child ages and so it is necessary to normalize these parameters for the effects of growth (Stansfield *et al.*, 2003). Hof (1996) recommended that temporal-distance parameters could be normalized by leg length, converting cadence and step length into dimensionless units as follows:

$$\begin{aligned}\text{dimensionless cadence} &= (\text{cadence}) / [g/(\text{leg length})]^{1/2} \\ \text{dimensionless step length} &= (\text{step length}) / (\text{leg length})\end{aligned}$$

The letter “g” in these equations represents the acceleration due to gravity (9.81 m/s<sup>2</sup>), leg length is measured in meters (the distance from the greater trochanter to the ground), cadence in steps per second and step length in meters (Hof, 1996). We were in possession of leg length measurements for the preoperative and 1, 3, 10 and 20 year postoperative studies and could apply this normalization method to our

measurements. This method enabled us to compare patients with themselves (over the 20 years of the study) and with the age-matched healthy controls.

### **Statistical Analysis**

Boxplots were used to graphically summarize the observed data. We performed 2 sets of comparisons: measurements at 1, 3, 10 and 20 years after SDR (the groups of interest) were compared with preoperative gait parameters and also with measurements from age-matched healthy control groups (the reference groups). For each gait parameter and comparison, we present the effect-size ( $\beta$ ), and the corresponding probability value. The  $\beta$  value reported is the difference between the means of the gait parameters (mean of the group of interest subtracted from the mean of the reference group), adjusted for age, sex and side as well as for repeated measures on each patient. The age we adjusted for is the age at time of operation (in the comparison with preoperative measurements) and current age (for comparison with healthy control groups). Individual linear mixed-effects models were used for all comparisons.

**Table 3.1** Characteristics of patients included in the 20 year follow-up study.

<i>Study</i>	<i>Group name</i>	<i>No. of Patients</i>	<i>Sex (F/M)</i>	<i>Mean Age in Yrs (range)</i>	<i>Source</i>
Preop	OCP	13	5/8	7.3 (2 - 14)	Vaughan <i>et al.</i> (1988, 1989)
1-yr postop	1CP	13	5/8	8.3 (3 - 15)	Vaughan <i>et al.</i> (1988, 1989)
	0-1 HC <sup>†</sup>	12	5/7	7.5 (2 - 12)	Vaughan <i>et al.</i> (1997)
3-yr postop	3CP	10	4/6	9.7 (5 - 17)	Vaughan <i>et al.</i> (1991)
	3 HC	12	5/7	9.2 (5 - 13)	Vaughan <i>et al.</i> (1997)
10-yr postop	10CP	10	4/6	16.1 (12 - 22)	Subramanian <i>et al.</i> (1998)
	10 HC	12	6/6	19.3 (14 - 22)	Subramanian <i>et al.</i> (1998)
20-yr postop	20 CP	13	5/8	27.3 (22 - 34)	Present study
	20 HC	12	5/7	27.8 (22 - 34)	Present study

HC = healthy control. <sup>†</sup> The same group of 12 healthy controls was used for the preoperative and 1 year postoperative comparisons.

## RESULTS

One of the 13 patients had conflicting feelings about the benefits of the SDR operation. All patients received intense physical therapy at school pre- and postoperatively, whereas more than half of the group also received orthopaedic surgery (tendon lengthening of rectus femoris, hamstring and Achilles, or osteotomy in the lower limb or foot) after SDR. Some of the patient cohort suffered from back pain, and all had relatively poor balance. Eleven of the 13 patients were either working or studying at college, and none had any problems with activities of daily living.

The results of this study are based on 13 patients with CP, of whom 3 were missing in the 3 and 10 year follow-up studies (Table 3.1). Table 3.2 shows the estimated adjusted differences between the subject groups and the reference groups ( $\beta$  values) and corresponding  $p$  values.

### ***Angular Kinematics***

#### *Knee and Hip Range of Motion*

Knee ROM of the patients with CP was, on average,  $15.9^\circ$  less than that of the healthy control group before surgery ( $p=0.002$ ). This ROM improved significantly ( $p<0.001$ ) at 1, 3, 10 and 20 years after the operation. The greatest difference ( $14.0^\circ$ ) was found 3 years after surgery. This mean was  $3.7^\circ$  greater than that of the healthy control group, although this difference was not significant ( $p=0.23$ , Table 3.2).

The mean hip range of motion was the only parameter that was not significantly different from the healthy control group before surgery. One year postoperatively it increased by 14.9 degrees ( $p<0.001$ ), and was 10.0 degrees greater than the healthy control group ( $p<0.001$ ). The data (Figure 3.1 and Table 3.2) show that after the initial large increase, the hip ROM values decreased and approached the healthy control values with no statistical differences.

### *Midrange Values*

The knee and hip midrange values of the patients with CP showed a similar trend in the years following surgery (Figure 3.2). Preoperatively, both parameters were greater in the CP group than in the healthy control group, although only the hip midrange value of  $9.5^\circ$  was significantly different ( $p=0.004$ ). One year postoperatively, there was a significant increase in both midrange values (knee:  $16.8^\circ$ ,  $p<0.001$ ; hip:  $4.0^\circ$ ,  $p=0.03$ ; Table 3.2). These values then improved, as indicated by a decrease in values closer to that of the healthy control groups (Figure 3.2), although they remained statistically greater (Table 3.2).

### *Temporal-Distance Parameters*

The dimensionless cadence of patients with CP did not change significantly after SDR, except at 20 years after surgery when there was a 0.07 mean increase compared with preoperative values ( $p=0.003$ ). Dimensionless cadence for patients was consistently less than that for healthy controls (Table 3.2, Figure 3.3).

The boxplots of dimensionless step length (Figure 3.3) and knee ROM (Figure 3.1) show a similar trends for patients with CP. One year after surgery there was a significant improvement of 0.13 ( $p<0.001$ ) compared with the preoperative value with a further significant increase (0.24) at 3 years ( $p<0.001$ ), followed by a slight deterioration at 10 and 20 years after surgery. Although the step length values of the 10 and 20 year follow-up studies are smaller than in the 3 year study and significantly different from the healthy control groups, there is still a significant benefit of 0.07 at 20 years after surgery compared to preoperative values ( $p=0.02$ , Table 3.2).

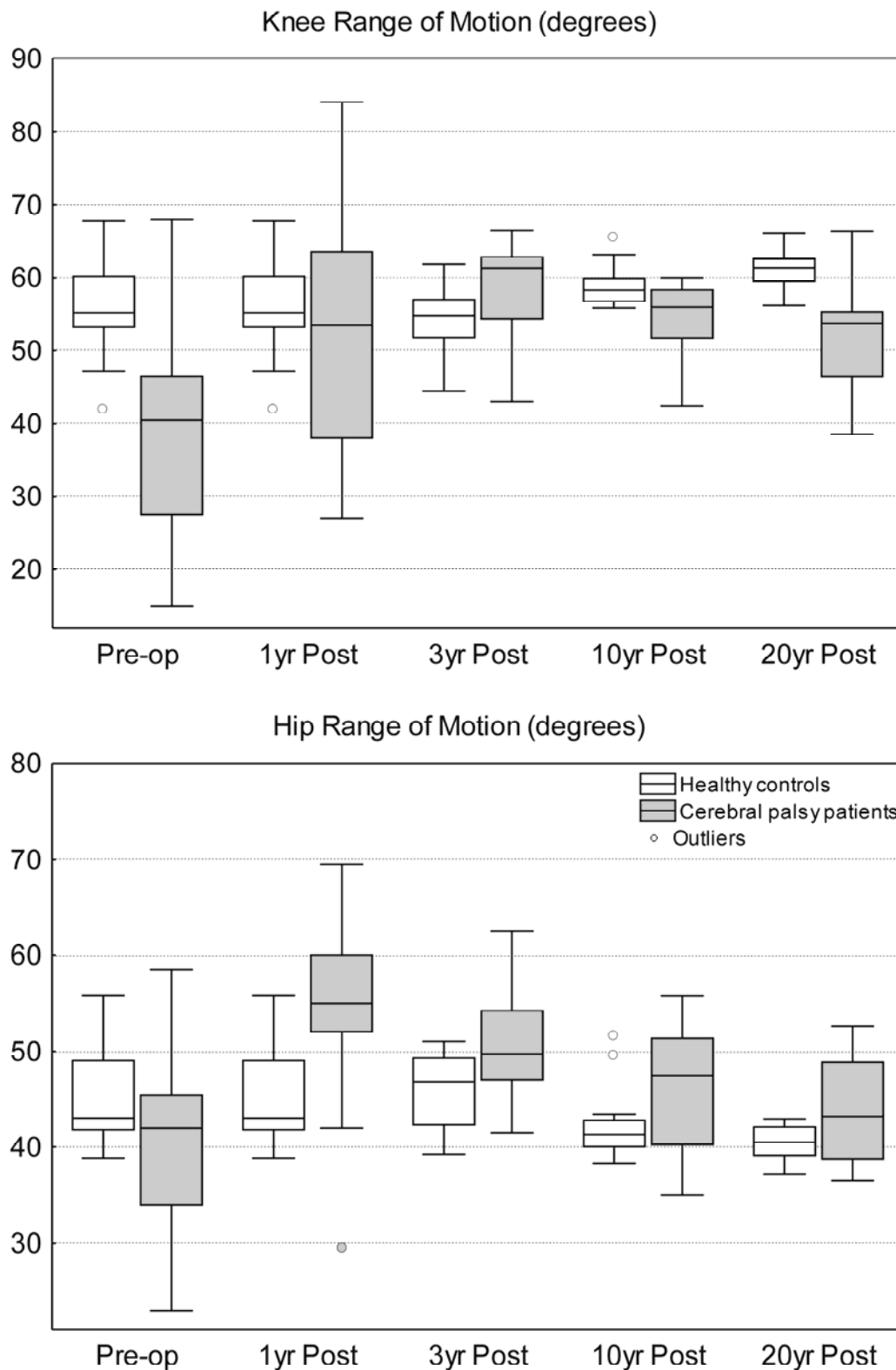
## **DISCUSSION**

Our study of patients with CP who underwent SDR in 1985 has shown that although the greatest gains were made during the first 3 years after surgery, 20 years later the patients still demonstrated an improvement in their gait pattern compared with preoperative values. This result is in accordance with our 10 year follow-up study, in which we objectively demonstrated the contribution of SDR to functional benefits, as measured by improved gait (Langerak *et al.*, 2007; Subramanian *et al.*, 1998).

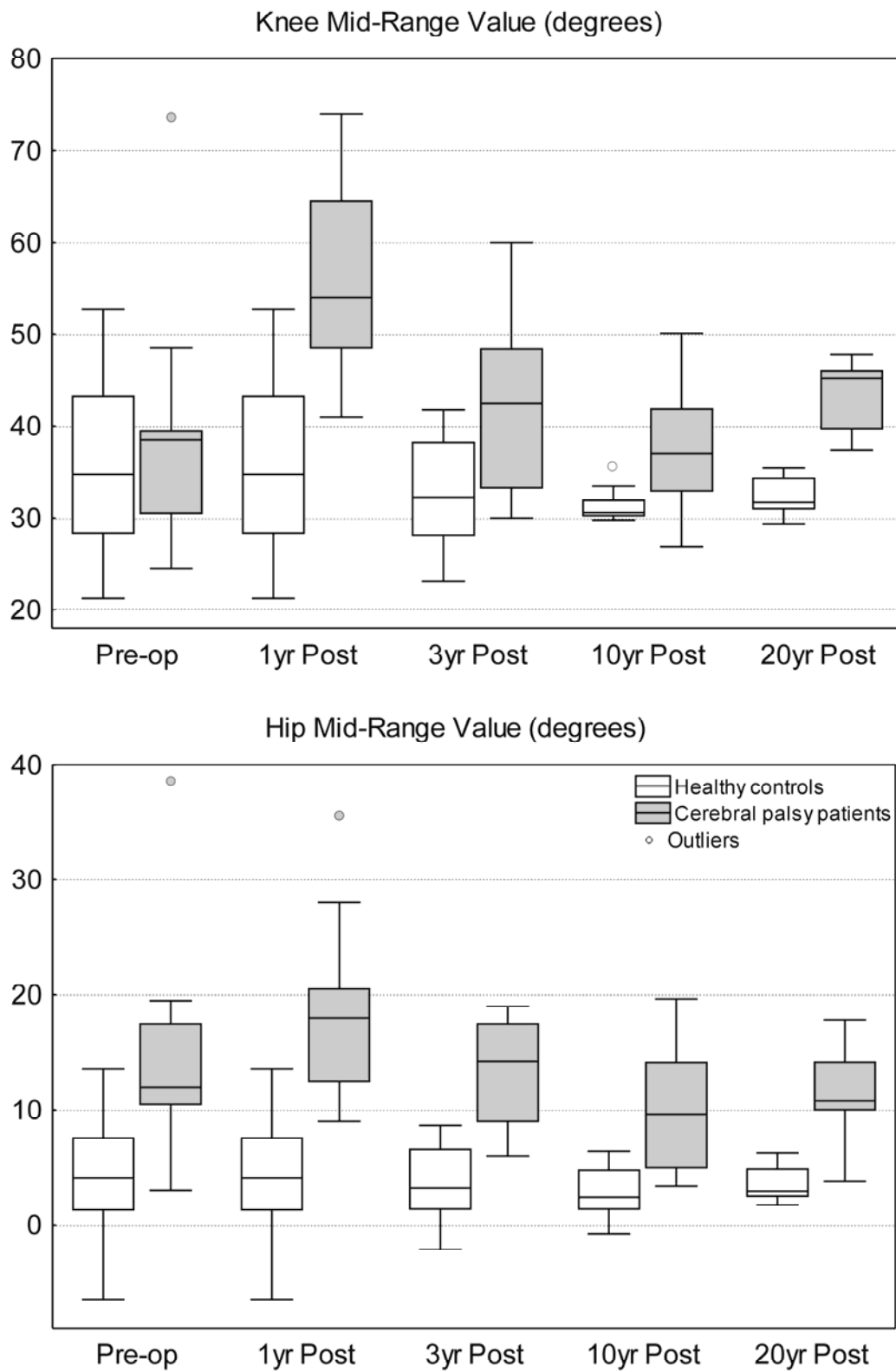
**Table 3.2** Adjusted differences in means and probability values for 6 gait parameters\*.

Gait parameter	Measures <sup>†</sup>	Preop (0CP) as Reference				Healthy Controls as Reference				
		1 CP	3 CP	10 CP	20 CP	0 CP	1 CP	3 CP	10 CP	20 CP
Knee ROM (°)	β	12.1	14.0	12.4	12.1	-15.9	-2.8	3.7	-4.0	-9.3
	p value	<0.001	<0.001	<0.001	<0.001	0.002	0.64	0.23	0.09	<0.001
Hip ROM (°)	β	14.9	8.6	4.6	3.9	-5.4	10.0	4.1	2.1	3.4
	p value	<0.001	0.003	0.01	0.04	0.07	<0.001	0.07	0.39	0.07
Knee midrange value (°)	β	16.8	4.1	-2.6	4.4	2.4	20.0	9.4	5.6	10.0
	p value	<0.001	0.21	0.41	0.07	0.55	<0.001	0.03	0.03	<0.001
Hip midrange value (°)	β	4.0	0.0	-4.0	-2.7	9.5	13.8	10.0	8.7	7.6
	p value	0.03	1.00	0.07	0.11	0.004	<0.001	<0.001	0.001	<0.001
Dimensionless cadence	β	-0.01	0.01	0.01	0.07	-0.09	-0.11	-0.07	-0.09	-0.06
	p value	0.61	0.72	0.73	0.003	0.04	0.002	0.06	0.02	0.005
Dimensionless step length	β	0.13	0.24	0.05	0.07	-0.24	-0.11	0.02	-0.18	-0.10
	p value	<0.001	<0.001	0.18	0.02	<0.001	0.09	0.58	0.001	0.002

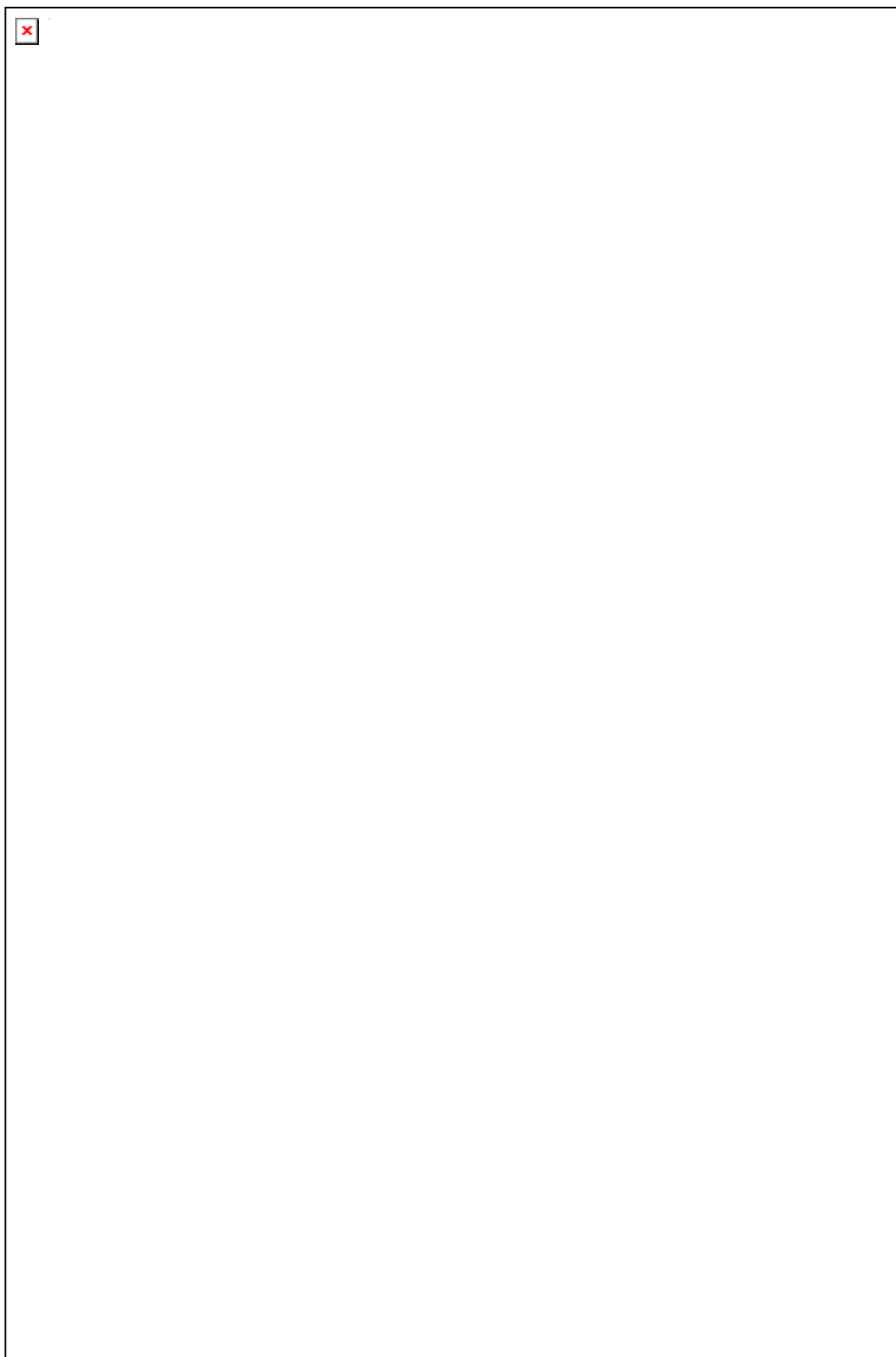
\* Adjusted for age, sex, side, and repeated measures. † β is calculated as the difference between the means of the gait parameters (mean of the group of interest subtracted from the mean of the reference group).



**Figure 3.1** Boxplot comparing knee (*upper*) and hip (*lower*) ROM values for patients with CP and healthy controls, before SDR (Preop) and at 1-, 3-, 10-, and 20 years postoperatively (Post). In all figures the *boxes* show the first, second (median), and third quartiles; *whiskers* represent the minimum and maximum values, excluding outliers; and *circles* represent the outliers.



**Figure 3.2** Boxplot comparing knee (*upper*) and hip (*lower*) midrange values for patients with CP and healthy controls, before SDR and at 1-, 3-, 10-, and 20 years postoperatively.



**Figure 3.3** Boxplot comparing dimensionless cadence (*upper*) and dimensionless step length (*lower*) values for patients with CP and healthy controls, before SDR and at 1-, 3-, 10-, and 20 years postoperatively.

### ***Angular Kinematics***

Patients with spastic CP demonstrate greater muscle stiffness than control groups (Damiano *et al.*, 2001). When these patients undergo an SDR, studies have shown that this spastic reduction will be maintained for at least 4 years (Gul *et al.*, 1999; Kim *et al.*, 2001; Mittal *et al.*, 2002; Peter and Arens, 1993). Diplegic patients have a smaller ROM than persons without spasticity (Wren *et al.*, 2005), and this will increase after SDR (Abel *et al.*, 2005; Damiano *et al.*, 2006). Our series of follow-up studies have shown this increase is most pronounced in the first 3 years after surgery (Langerak *et al.*, 2007; Subramanian *et al.*, 1998; Vaughan *et al.*, 1991), during the important years of growth and may lead indirectly to fewer contractures. As the patients became older, our 10 and 20 year follow-up studies showed slightly decreased ROM for the hip and knee (Figure 3.1).

The typical walking pattern for patient with CP and spastic diplegia is a crouch gait, which results in a high midrange value for the hip. One year after SDR, a “collapsed” gait pattern is observed, with a significant increase in midrange values for both knee and hip (Figure 3.2). One reason for this negative side-effect could be postoperative muscle weakness in the lower extremities postoperatively. Alternatively, we hypothesize that this change in gait mechanics could also be the result of lumbar hyperlordosis, which leads to increased hip flexion to maintain a patient’s center of gravity over the base of support.

### ***Temporal-Distance Parameters***

Damiano and Abel (1998) showed that cadence is correlated with muscle strength in the lower extremities. In general, muscle strength in patients with CP is less than that of healthy controls, which may be why the number of steps per minute is reduced. The outlier in dimensionless cadence represented a single patient (Figure 3.3). She and her mother reported in the questionnaire that, although she received intensive physical therapy postoperatively, she continued to suffer from muscle weakness. Although her cadence increased slightly at 10 and 20 years after surgery, this increase was probably not a result of increased muscle strength but rather because she was walking with a crutch.

In contrast to cadence, step length is not correlated with muscle strength (Vaughan *et al.*, 1989), but is related to joint excursion, which is reduced in patients with CP compared with healthy controls (Abel *et al.*, 2005; Damiano *et al.*, 2006). In our 20 year follow-up study, the changes in knee ROM corresponded with the changes in dimensionless step length (Figures 3.1 and 3.3).

### ***Gait in Adults with Cerebral Palsy***

Andersson and Mattsson. (2001) and Jahnsen *et al.* (2004) have published questionnaire data regarding the locomotion skills of adults with CP. Their groups consisted of 77 and 145 patients with diplegic CP, respectively, and they showed that 30 to 40% of the patients were able to walk without support, although 20% always used a wheelchair. Both studies showed that 75% of their patients demonstrated decreased walking ability between 15 and 34 years of age. Although we have to consider that our group size was much smaller (14 patients), we demonstrated that 20 years after SDR, at a mean age of 27.3 years, 11 of our patients (79%) walked without support and only 1 (7%) was confined to a wheelchair.

### ***Limiting Factors***

Fourteen patients with CP participated in our studies was 14, which provided sufficient power for the statistical analysis; however, the research sample should ideally be much larger for our findings to have widespread applicability. In addition, 3 of the 14 patients were missing for the 3 and 10 year follow-up studies, and we had to exclude 1 of the 14 because she was not able to walk independently 20 years after surgery. Because of the reduced sample size, we were unable to account for confounding factors such as orthopaedic surgery and back pain.

The healthy control groups for the various postoperative studies did not consist of the same study participants, whereas the CP patient group did. In our analysis, we used the appropriate statistical methods to factor in this discrepancy. Ideally, we should also have had a single control group consisting of patients with CP who did not undergo SDR in 1985. This was not ethically possible, however, and for that reason we used age-matched healthy persons. There is also a dearth of published gait data for adults with CP, making it difficult to compare our patients at 20 years after surgery.

## CONCLUSIONS

Since new information on SDR was obtained in Cape Town in the early 1980's, few centers have had the opportunity to perform long-term studies. We conclude that 20 years after our patients underwent SDR, they continue to show an improvement in their gait patterns compared with preoperative levels. Although we cannot predict how these patterns might have looked without this operative procedure, we can assume that the patients would continue to have experienced spasticity and deterioration from secondary abnormalities such as contractures. In addition to the positive locomotor benefits of SDR 20 years after surgery, some concerns remain about spinal instability (Peter *et al.*, 1993). We believe it is necessary to conduct detailed research about these factors, thus affirming the importance of long term clinical follow-up studies.

## ACKNOWLEDGEMENTS

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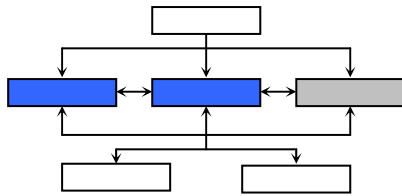
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# Chapter 4



Functional status of patients with cerebral palsy according to the international classification of functioning, disability and health model: a 20 year follow-up study after selective dorsal rhizotomy

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## ABSTRACT

**Objective:** To determine functional status of patients with cerebral palsy 20 years after they received selective dorsal rhizotomy (SDR). **Design:** A prospective 20 year follow-up study. **Setting:** Red Cross Children's Hospital (SDR operation and 1 year follow-up assessment) and at institutional or private locations nearby patients' homes (20 year follow-up assessment). **Participants:** Referred sample of 14 patients with spastic diplegia (6 women, 8 men; mean age, 27y; range, 22–33y) who were preoperatively ambulant and fulfilled strict selection criteria for SDR operation in 1985. **Interventions:** Patients were assessed before and 1 and 20 years after SDR. **Main Outcome Measures:** Standardized assessments of function according to 2 dimensions of the *International Classification of Functioning, Disability and Health* (ICF) model: (i) *Body Function & Structure* (muscle tone, joint stiffness, voluntary movement) and (ii) *Activity* (rolling, sitting, kneeling, crawling, standing, walking, transitions) were obtained. In addition, based on assessments and questionnaires, Gross Motor Function Classification System (GMFCS) levels were determined before and at 1 year after SDR retrospectively and currently at 20 years after SDR. **Results:** One year after SDR, functional outcomes based on the 2 dimensions of the ICF-model improved significantly, and these improvements were maintained at 20 years after surgery. Patients showed a shift in their GMFCS levels 1 and 20 years after SDR. **Conclusions:** In line with our 20 year follow-up study with gait parameters as outcome measures, patients with spastic diplegia still show improvements in their functional status 20 years after SDR. We acknowledge the presence of possible confounding factors and a small sample size, but we argue that the improvements found in this study were caused mainly by SDR. Finally, changes in GMFCS levels suggest a possible role for this tool to detect changes after an intervention.

**Key Words:** Adult • Cerebral palsy • Follow-up studies • Rehabilitation • Rhizotomy • Spastic diplegia

## INTRODUCTION

During the past few decades, the prevalence of cerebral palsy (CP) has risen in developed countries from 1.5 to 2.5 per 1000 live births, and these patients now constitute the largest diagnostic group in paediatric rehabilitation (Odding *et al.*, 2006). About 80 percent of these children are diagnosed with spasticity (Odding *et al.*, 2006; Stanley *et al.*, 2000), a condition which may disturb the normal growth process and can lead to secondary abnormalities such as deformities and pain (Gage, 2004; Liptak, 2008; Andersson and Mattsson, 2001; Hilberink *et al.*, 2007; Jahnsen *et al.*, 2004). Therefore, it is important to treat spasticity in these children to facilitate their optimal physical development.

One of the possible treatment options to reduce spasticity in children with CP is selective dorsal rhizotomy (SDR). In the early 1980's Peacock and Eastman (1981) reintroduced and refined this neurosurgical procedure in Cape Town, South Africa. During this operation laminectomies are performed from L2 to S1, which exposes the cauda equina. After separating the dorsal roots from the anterior roots, the dorsal roots are electronically stimulated and the electromyographic (EMG) response of the corresponding muscle is recorded. Those dorsal nerve rootlets associated with a normal EMG response are left intact, whereas those associated with an abnormal response are divided. By transecting a percentage of dorsal lumbar sacral rootlets, the myotactic reflex arc is disrupted which decreases muscle spasticity in the lower extremity.

Because the manifestation of CP and spasticity is complex, the International Classification of Functioning, Disability and Health (ICF) model, which was developed by the World Health Organisation (WHO) in 2001 (WHO, 2001), can be used to quantify the outcomes and determinants of CP (BurrIDGE *et al.*, 2005; Rosenbaum and Stewart, 2004; Majnemer and Mazer, 2004; Liptak, 2008). Steinbok published an extensive review of 63 articles that examined the outcomes of SDR in 2001, and reanalyzed his findings in 2007 in the context of the ICF-model (2001; 2007). All these studies supported the finding that SDR decreases spasticity and increases joint range of motion in the lower extremity (ICF-model dimension: *(i) Body Function & Structure*). Strong, although inconclusive, evidence was found for improvements in

motor function (ICF-model dimension: *(ii) Activity*), whereas only moderate evidence was found for a reduction in level of disability (ICF-model dimension: *(iii) Participation*) (Steinbok, 2001; Steinbok, 2007).

Although these findings are promising, most conclusions are based on follow-up studies not longer than 5 years after SDR. Because the refined SDR operations were first performed in Cape Town, we are in the unique position of being able to provide insight into the long-term sequelae of this neurosurgical procedure (Langerak *et al.*, 2007). As a result, we recently published a 20 year follow-up study that showed the long-term benefits of SDR in locomotor function (Langerak *et al.*, 2008). Although gait analysis is a reliable and objective research method, there is nevertheless a demand for more detailed long-term functional outcomes after SDR. In an attempt to provide a complete picture of these patients' function, we conducted, in line with our 1 year follow-up study (Berman, 1989; Berman *et al.*, 1990), functional assessments 20 years after SDR. Accordingly, the aim of this study was to determine the functional status based on the following ICF-model dimensions: (i) *Body Function & Structure* and (ii) *Activity* in a group of patients with spastic diplegia who had already shown a continued improvement in their gait pattern 20 years after SDR.

## METHODS

### *Participants*

The selection of participants was based on the following criteria: (1) diagnosed with spastic diplegia of congenital origin; (2) fulfilled strict selection criteria for SDR; (3) received SDR by a single neurosurgeon (WJP) at the Red Cross Children's Hospital in Cape Town (South Africa) in 1985; (4) had access to intensive physiotherapy before and after surgery; and (e) ambulant preoperatively.

Fourteen patients met the inclusion criteria and had participated in the original 1 year follow-up study (Berman, 1989; Berman *et al.*, 1990). After an extensive search because of the absence of contact details and a non-existing national CP registration system, we tracked down and contacted each of the 14 patients in 2005. Before entering the study, the aim and purpose of the research project was clearly explained

to each patient, after which an informed consent was signed. This study was approved by the Human Ethics and Research Committee of the Faculty of Health Sciences of the University of Cape Town.

### **Procedures**

Preoperatively in 1985, 2 days prior to surgery, a single occupational therapist assessed the participants' neuromuscular and functional status in a clinical room at the Red Cross Children's Hospital. In 1986, on average 1 year after surgery, the same investigator conducted the re-assessments of each child using the same protocol (Berman, 1989; Berman *et al.*, 1990).

Because the SDR operation was performed on patients from all over South Africa, 8 of the 14 participants were not living in or close to Cape Town 20 years after SDR (in 2005). These patients were visited and tested either at home or at a suitable location nearby. Neuromuscular and functional assessments were repeated by a single physiotherapist (NGL) using the original protocol (Berman, 1989). A second physiotherapist (RPL), who recorded all assessments on video tape, was consulted when a clear judgement could not be made by the principal researcher (NGL). After the assessment the video tape of each patient was reviewed by the two experienced physiotherapists (NGL and RPL) to confirm the final scores chosen. The two therapists were blinded from any neuromuscular and functional outcomes of previous studies during this process.

### **Assessment Tools**

#### *International Classification of Functioning, Disability and Health model: Body Function & Structure*

In line with the original 1 year follow-up study, an ordinal assessment scale was used to quantify outcomes of the first dimension of the ICF-model, *Body Function & Structure*, which included the following items (Berman, 1989): 16 for muscle tone, the limb was moved passively in a direction opposite to the action of the muscle group being tested, and the resistance to passive stretch was noted. This outcome measure is comparable to the Modified Ashworth Scale. However, outcomes of the muscle tone parameter were not identical and so it was not possible to convert to this worldwide accepted assessment tool. In Appendix I, the defined scores for muscle

## Chapter 4

tone are presented in detail (scale from low [1] and normal [2] to most severe increase [5] in muscle tone).

To measure joint stiffness, each lower-limb joint was tested passively by moving the limb through the maximum range of movement and compared with the full range of joint movement in a normal limb. The score for this parameter was based on the maximum passive range of motion achieved, taking into account the manner wherein this movement was obtained. The score for joint stiffness varied from normal [1] to total limitation like an apparent fixed contracture [4] (Appendix I).

For voluntary movement measurement, the patient was asked to stop and start a movement 8 times within his/her full available range of movement, counting with each movement. The patient was positioned to eliminate the effects of gravity. The joints proximal to the movement were stabilized in a position to inhibit mass patterns as far as possible.

Outcomes of voluntary movement consist of a combined assessment of selective motor control, muscle strength, and joint range of motion (scale from normal [1] to most severe limitation [5] [see Appendix I]).

Muscle tone, joint stiffness, and voluntary movement assessments were measured for the following 10 movements: hip flexion, extension, abduction, and adduction; knee flexion and extension; and ankle inversion, eversion, plantar, and dorsiflexion. To quantify the total outcome of each item optimally, a mean score for all 10 movements was calculated for both the right and left legs.

### *International Classification of Functioning, Disability and Health model: Activity.*

To quantify the functional outcomes according to the second dimension of the ICF-model, *Activity*, the following item of the original ordinal assessment scale (Berman, 1989) was used: the quality of functional movement was tested by asking the patient to assume and maintain various positions including rolling, long and side sitting, prone and half kneeling, kneel standing, crawling, standing, and walking. This assessment tool bears some resemblance to the wellknown Gross Motor Function Measure (GMFM), which was developed in the early 1990s after our 1 year follow-up

study. Therefore, we did not apply the GMFM outcome measures, but rather we used the comparable functional movement assessment tool as described by Berman.<sup>16</sup> (scale from normal [1] to most severe limitation [5] [Appendix II]).

Each of these developmental movements allows researchers and clinicians to quantify a patient's ability to dissociate and align his/her body. Therefore, the outcomes of the functional movements were determined separately and as an overall mean for all 9 movements.

#### *Questionnaire and Gross Motor Function Classification System*

In addition to the primary functional assessments, a questionnaire was completed with the following topics: medical interventions received, current medical issues, use of assistive devices, employment and/or student status, and problems with daily activities.

Based on the current questionnaire and the assessment outcomes, each participant was classified into the Gross Motor Function Classification System (GMFCS) in 2005 (Palisano *et al.*, 1997; Jahnsen *et al.*, 2006). Although retrospective classification into GMFCS levels has not yet been validated, we used a detailed questionnaire plus the original clinical reports and the functional assessments from 1985 and 1986 to assign GMFCS levels for our 14 participants before and 1 year after SDR.

The GMFCS categorizes patients with CP, based on their gross motor performance, into levels ranging from level I (minor limitations) to level V (severe limitations) (Palisano *et al.*, 1997). Although it is debatable whether the GMFCS can be used to measure changes over time (Rosenbaum *et al.*, 2008), we decided to use this classification system to show a possible shift in gross motor function after SDR, as was done by Cole *et al.* (2007).

#### **Statistical Analysis**

We used the nonparametric Friedman test to determine the influence of SDR over time for each of the functional outcome measures. If statistical significance between the preoperative and postoperative studies was reached, we used the Wilcoxon matched-pairs test to determine where the significant difference appeared. Because

the outcome measures were analyzed at 3 different time points (preoperatively and at 1 and 20 years postoperatively), this analysis resulted in 3 different paired comparisons. Bonferroni correction for multiple comparisons was used, which resulted in a  $p$  value of  $0.05/3$  equal to  $0.017$  that was used to determine statistical significance (Bland and Altman, 1995).

## RESULTS

### ***Characteristics of Participants***

The mean age of the 14 patients with spastic diplegia was  $28 \pm 4$  years (range, 22 - 33y) 20 years after SDR. The cohort consisted of 6 women and 8 men from all socioeconomic backgrounds and races. Current GMFCS levels in the participants ranged from GMFCS level I ( $n=7$ ), level II ( $n=3$ ), level III ( $n=3$ ), to level IV ( $n=1$ ). The cognitive function of the participants before SDR varied from moderate ( $n=3$ ) to mild ( $n=11$ ) impairment. All participants attended a specialized school for children with CP and received intense physiotherapy (and if necessary occupational therapy) before and after surgery.

### ***International Classification of Functioning, Disability and Health Model: Body Function & Structure***

Muscle tone, joint stiffness, and voluntary movement scores all showed a significant difference when comparing before and after SDR (Friedman test,  $p < 0.001$ ). As shown in the box plots (Figure 4.1), the scores for these parameters improved as represented by a decrease in each score. The median score for muscle tone was significantly reduced from 3.1 to 2.1 at 1 year after surgery and to 2.0 at 20 years after SDR, which is equivalent to normal muscle tone (Table 4.1, Figure 4.1a). In addition, the median joint stiffness score was significantly reduced, from 1.9 to 1.2, at 1 year after surgery, and this improvement was maintained with a median score of 1.3 at 20 years after surgery (Table 4.1, Figure 4.1b). The voluntary movement score was significantly decreased from a median score of 3.6 before surgery to 2.3 at 1 year after surgery and decreased further, after 20 years, to a median score of 1.9, which approached a normal score of 1.0 (Table 4.1, Figure 4.1c). As presented in Table 4.1, none of the outcome measures of the ICF-model dimension *Body Function*

& Structure showed a significantly different score between 1 and 20 years after SDR. Note that the highest scores for muscle tone (2.8), joint stiffness (3.4), and voluntary movement (3.6) 20 years after SDR represented a single participant who was classified at GMFCS level IV.

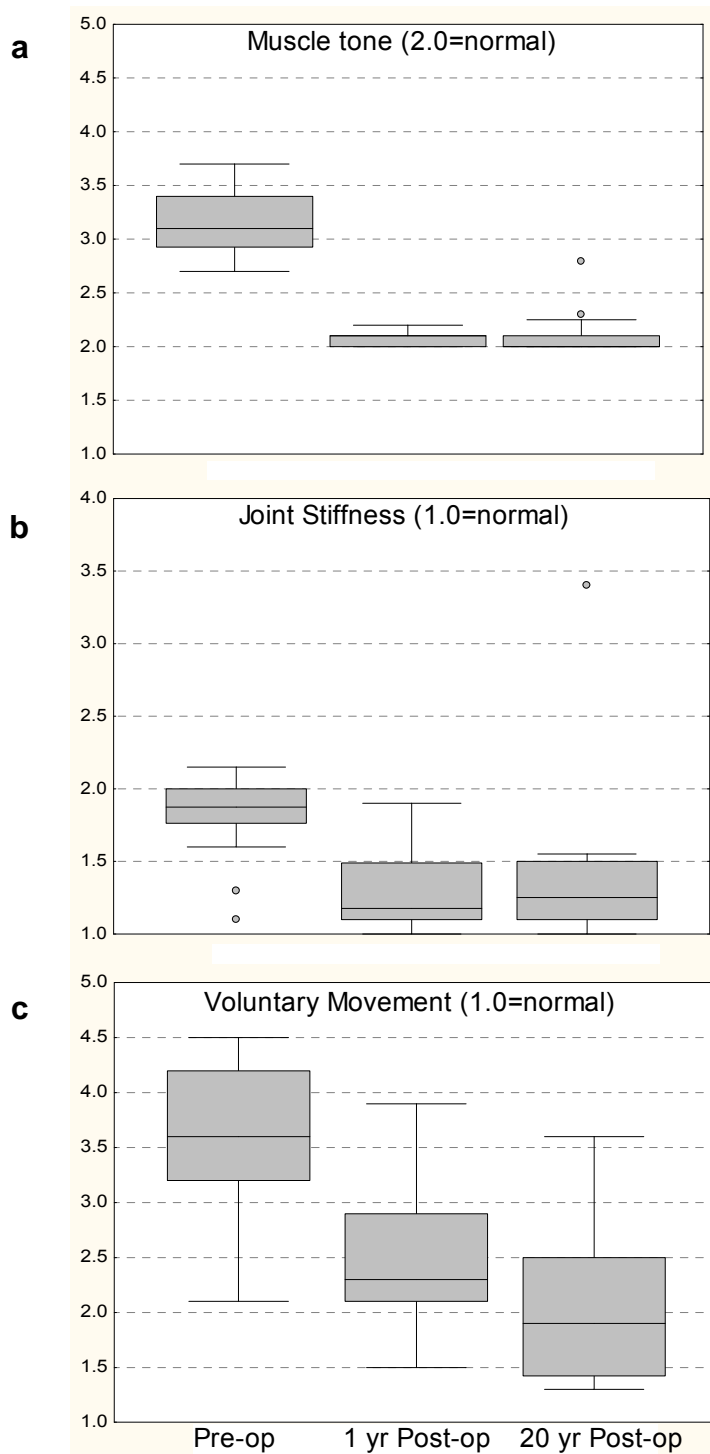
**Table 4.1** *P* values from Wilcoxon matched-pairs test tabulated for scaled scores of neuromuscular and functional Outcomes.

<i>Outcome measure</i>	<i>p value</i>		
	<i>Pre-1yr</i>	<i>Pre-20yr</i>	<i>1y-20yr</i>
Muscle Tone	<0.001*	<0.001*	0.859
Joint Stiffness	0.001*	0.019	0.972
Voluntary Movement	0.001*	0.002*	0.021
Functional Movement	<0.001*	<0.001*	0.328
Long sitting	0.001*	0.001*	0.074
Side sitting	0.005*	0.074	0.937
Prone kneeling	0.008*	0.005*	0.361
Kneel standing	0.001*	0.002*	0.463
Half kneeling	0.001*	0.003*	0.024
Standing	0.012*	0.028	1.000
Rolling	0.005*	0.004*	0.463
Crawling	0.028	0.008*	0.142
Walking	0.008*	0.050	0.208

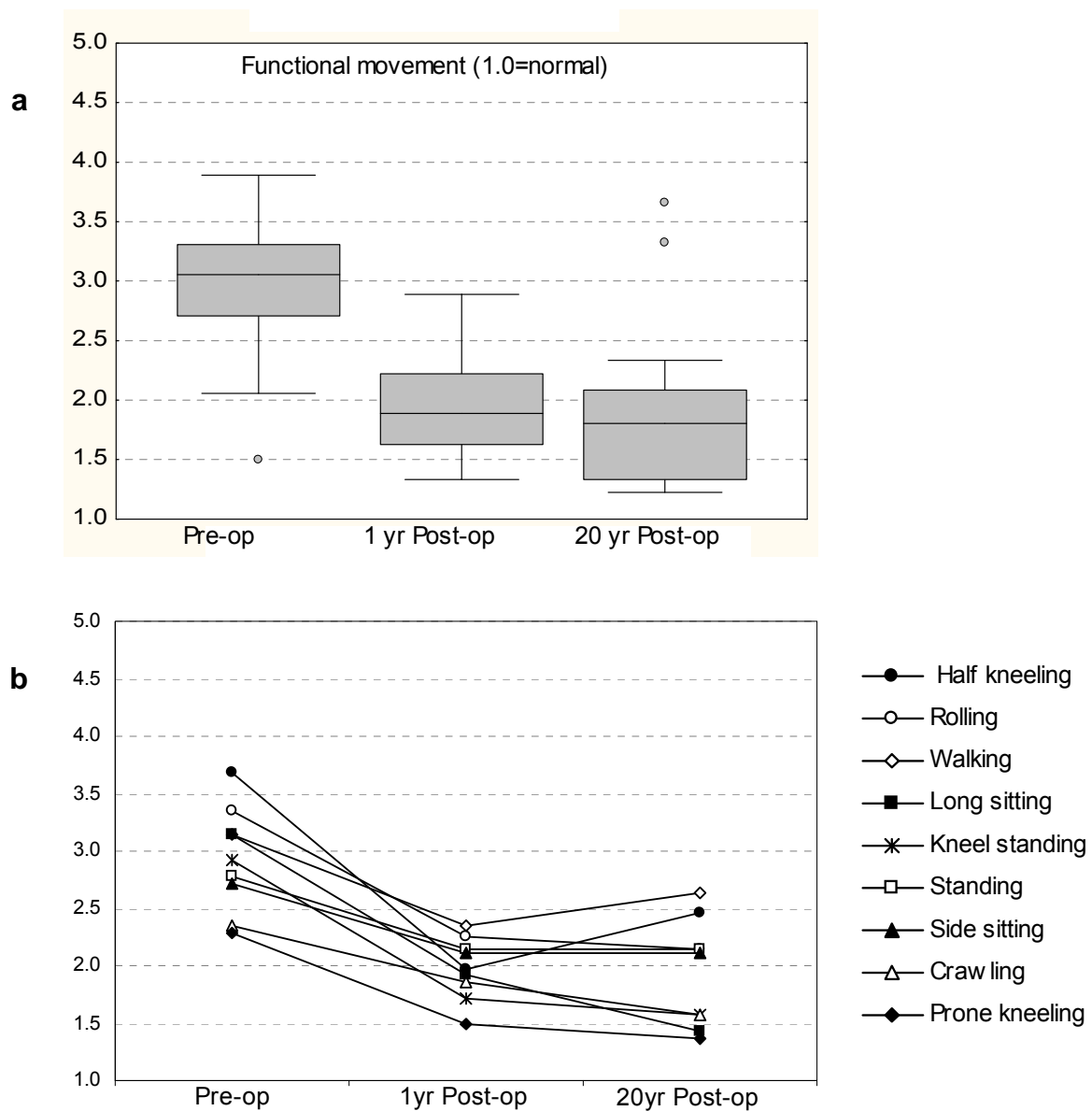
\*Significant difference achieved by a *p* value of 0.05/3 = 0.017. Abbreviations: Pre = preoperative; 1yr = 1 year postoperative; 20yr = 20 year postoperative.

### ***International Classification of Functioning, Disability and Health Model: Activity***

The mean functional movements scores and the 9 separate assessment scores all showed a statistical effect in time (Friedman test,  $p < 0.001$ ). The box plots show an improvement with a reduced score approaching the normal level of 1.0 at 1 and 20 years after surgery (Figure 4.2). The preoperative scores decreased significantly from 3.1 to 1.9 and 1.8 at 1 and 20 years after surgery, respectively (Table 4.1). Figure 4.2 provides an overview of the median scores of the 9 different assessments before and after SDR. Note that the outlier with a mean functional movement score of 3.7 represented the participant who was classified at GMFCS level IV 20 years after SDR.

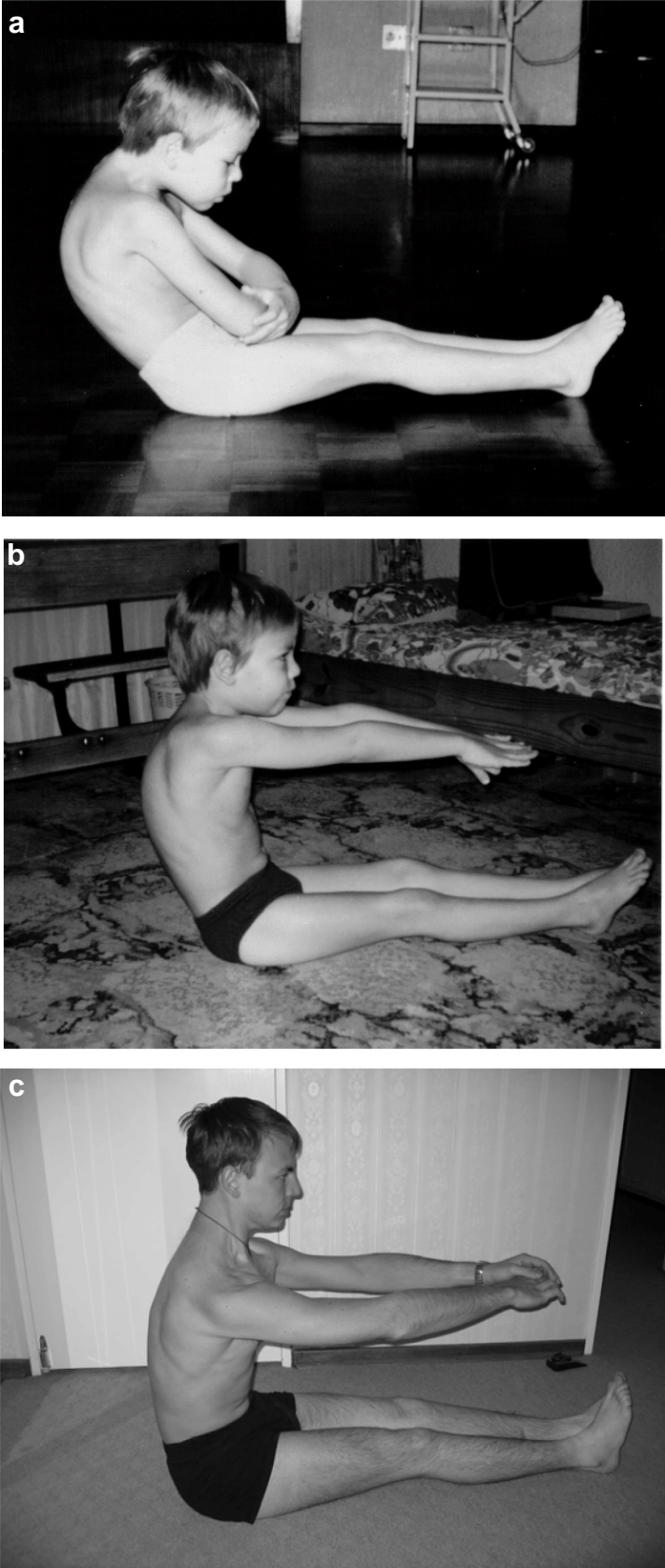


**Figure 4.1** Outcome measures for the ICF dimension *Body Function & Structure* before and at 1 and 20 years after surgery. Box plots show (a) muscle tone; (b) joint stiffness; and (c) voluntary movement, whereas whiskers represent the minimum and maximum values, excluding outliers. Outliers are values more than 1.5 box lengths from the upper or lower edge of the box and shown as circles. Note: the median of muscle tone scores at 1 and 20 years postoperatively are 2.1 and 2.0, respectively.



**Figure 4.2** Outcome measures for the ICF dimension *Activity* before and after surgery. (a) Box plots show 1<sup>st</sup>, 2<sup>nd</sup> (median), and 3<sup>rd</sup> quartiles, whereas whiskers represent the minimum and maximum values, excluding outliers. Outliers are values more than 1.5 box lengths from the upper or lower edge of the box and are shown as circles. (b) Line plots of median scores of 9 functional movements before and at 1 and 20 years after surgery.

The assessment of long sitting, prone kneeling, kneel standing, half kneeling, and rolling resulted in significant improvements at 1 year and 20 years after surgery (Table 4.1). The median score for crawling at 1 year after surgery was reduced compared with preoperative values, although not significantly. Twenty years after SDR, the score was further decreased, which resulted in a significant difference when compared with preoperative values. Side sitting, standing, and walking assessments resulted in a significantly different score 1 year after surgery, but 20



**Figure 4.3** Patient with CP showing 1 of the functional movement outcome measures: long sitting before surgery (a), at 1 year (b), 20 years (c) after surgery (Berman *et al.*, 1990).

years later the score was not significantly improved when compared with the values before dorsal rhizotomy. None of the 9 movement scores changed significantly between 1 and 20 years after SDR (Table 4.1).

Figure 4.3 shows one of the participants demonstrating long sitting before surgery and at 1 year and 20 years after surgery. These images show improved hip flexion and knee extension and better control of the head and trunk after dorsal rhizotomy.

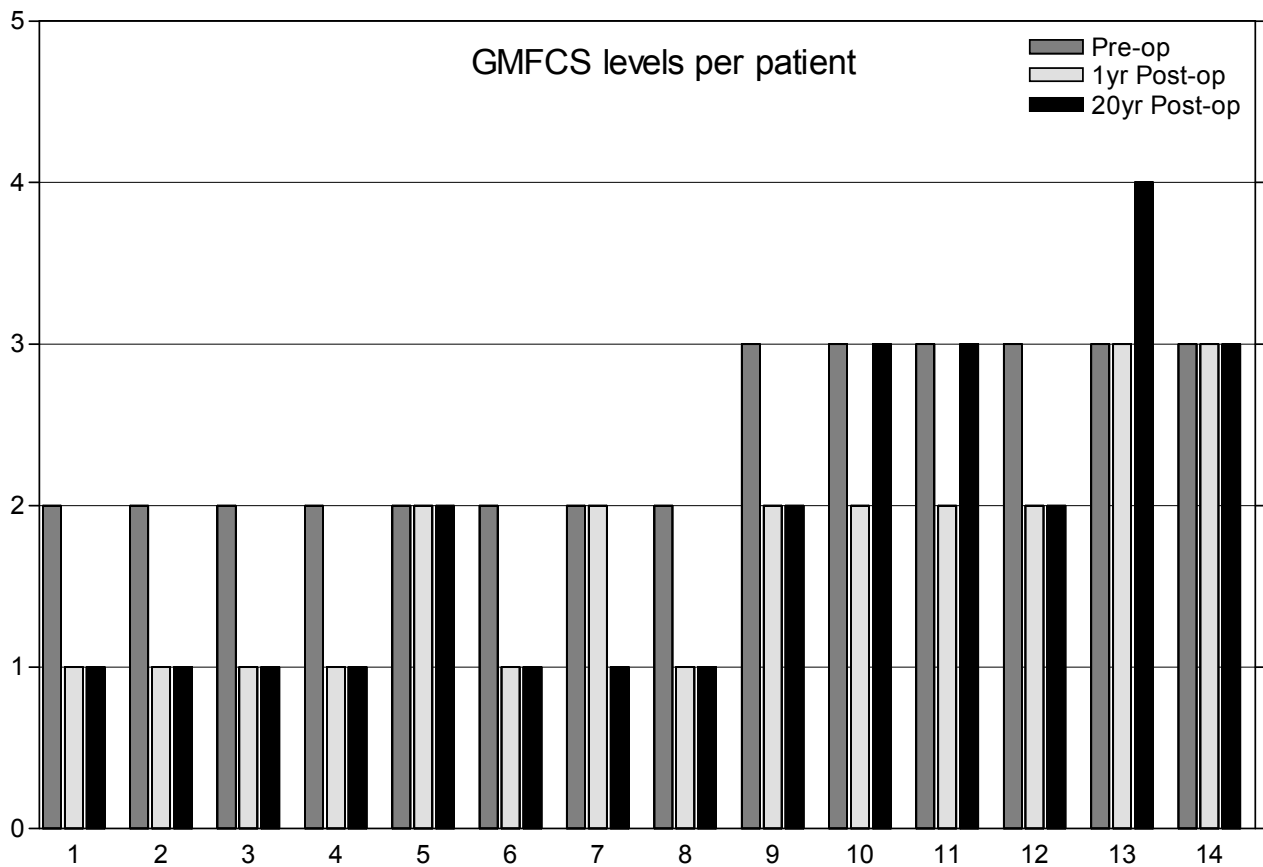
### ***Questionnaire and Gross Motor Function Classification System***

After surgery, 9 participants received at least 1 orthopaedic surgery procedure, including muscle releases (Achilles' tendon: n=2, hamstring: n=3, rectus femoris: n=1) and/or osteotomies (foot: n=6, femur: n=1). However, none of them received an intrathecal baclofen pump or botulinum toxin injections, and only 1 participant (classified at GMFCS level IV) occasionally used oral antispasmodic medication after SDR.

Twenty years after SDR, 11 participants walked in- and outdoors independently, whereas 2 participants walked with a cane or crutch, and only 1 participant (classified at GMFCS level IV) became a wheelchair user. All participants reported problems with holding their balance, whereas the majority (n=10) experienced regular back pain and 1 person had a bladder dysfunction (which was controlled by medication). However, all participants answered "no" to the question "Do you need help with activities of daily living?" Eleven of the 14 participants were currently employed or were studying at college, whereas 5 actively participated in physical exercise.

Figure 4.4 provides an overview of the GMFCS levels of the 14 participants before and at 1 and 20 years after SDR. One year after surgery, 10 participants were classified at a lower GMFCS level (functional improvement), whereas 4 remained at the same level. Between 1 year and 20 years after SDR, 2 participants deteriorated from level II to III, 1 participant deteriorated from level III to IV and 1 participant improved from level II to I. In 2005, 20 years after surgery, 9 participants were still classified in a lower GMFCS level compared with their levels before SDR, whereas 4

participants remained unchanged and 1 participant deteriorated from GMFCS level III to IV.



**Figure 4.4** Distribution of GMFCS levels per patient (n=14) before and at 1 and 20 years after SDR

## DISCUSSION

A modification of SDR was introduced in Cape Town in the early 1980s (Peacock and Eastman, 1981), and became widespread after the relocation of Warwick Peacock from South Africa to Los Angeles in 1986. During the past 2 decades, dorsal rhizotomy has become an accepted method to release children with CP from their spasticity, and it has produced a positive influence in patients' function, activities, and participation in daily living during the first few years after surgery (Steinbok, 2001; 2007). Besides our 20 year follow-up studies, no other studies have been performed to determine the long-term effects of SDR on patients with CP. In line with our gait study (Langerak *et al.*, 2008),<sup>15</sup> outcomes of functional

assessments based on the 2 dimensions of the ICF-model, *Body Function & Structure* and *Activity*, showed improved values 20 years after SDR compared with preoperative values.

### ***Outcomes Related to the 3 Dimensions of the International Classification of Functioning, Disability and Health Model***

With regards to the first dimension of the ICF-model, *Body Function & Structure*, numerous studies have shown improvements in neuromuscular outcome measures after SDR. Steinbok (2001) concluded in his review article that dorsal rhizotomy results in a significant reduction in muscle tone in the first few years after surgery. Short-term follow-up studies, up to 5 years after SDR, by Gul *et al.* (1999) and Mittal *et al.* (2002a) have shown improvements in muscle tone. In line with these outcomes, the present study shows a significant reduction in muscle tone 1 year after dorsal rhizotomy, which was maintained 20 years after SDR (Figure 4.1a).

When spasticity is not treated properly, it can lead to secondary abnormalities such as muscle contractures and decreased range of motion (Gage, 2004). Because dorsal rhizotomy reduces muscle tone in the lower limbs, it also improves the range of motion in these joints (Steinbok, 2001). Gul *et al.* (1999) and Mittal *et al.* (2002a) showed an augmented or at least a stabilized range of motion in lower-limb joints between 1 and 5 years after SDR. Our Cape Town research group, which measured range of motion based on gait analysis, found improvements in range of motion up to 20 years after SDR (Vaughan *et al.*, 1988; Vaughan *et al.*, 1991; Subramanian *et al.*, 1998; Langerak *et al.*, 2008). This latter finding is in line with the current study, which found a decreased joint stiffness (a measure of range of motion during passive movement) 1 year after SDR that was maintained at 20 years after surgery (Figure 4.1b). This long-term result would appear to be important because a descriptive study in adults with CP who did not receive SDR showed that 77% of a cohort of 77 with spastic diplegia reported contractures in their extremities (Andersson and Mattsson, 2001).

Voluntary movement, which was assessed in this study, measured a combination of range of motion, motor control, and muscle strength. Steinbok's review article (2001) showed either an unchanged or increased muscle strength after SDR. A shortterm

follow-up study by Gul *et al.* (1999) reported improved muscle strength in hip abductors, dorsiflexors, and hip extensors between 1 and 5 years after SDR in which the muscle strength of the quadriceps remained the same. These findings are in line with the initial and further improvements found in voluntary movements at 1 and 20 years after SDR (Fig 4.1c).

Related to the second dimension of the ICF-model, *Activity*, changes in functional movements were measured after SDR. Steinbok (2001) concluded in his review article that there is strong, although inconclusive, evidence that dorsal rhizotomy also leads to benefits in functional outcome measures. Short-term follow-up studies showed a significant improvement in GMFM (Mittal *et al.*, 2002a) and functional tasks and positions (Mittal *et al.*, 2002a; Gul *et al.*, 1999) 1 year after SDR, with a further improvement 5 years after surgery. This current study shows that the improvements in 9 functional movements gained 1 year after SDR are maintained at 20 years (Figure 4.2).

A third dimension of the ICF-model, *Participation*, could unfortunately not be quantified in this study. The original questionnaire (used before and 1 year after SDR) only included a limited amount of questions, and so we were not able to get a complete and accurate picture of this dimension. Steinbok (2001), however, reported improvements in participation measured through the Pediatric Evaluation of Disability Inventory and FIM for Children after SDR. Mittal *et al.* (2002b) found improvements in participation measured by the Pediatric Evaluation of Disability Inventory 3 and 5 years after SDR. Based on our questionnaire, which was completed in 2005, we noted that none of our 14 patients reported problems with daily activities 20 years after SDR. In addition, we found that 28% of our cohort actively participated in physical training, whereas 86% were employed and 14% were studying. In contrast, Andersson and Mattsson (2001) reported higher rates of physical training (66%), whereas a lower percentage of employment and study rates were found (47% - 64% and 9%, respectively) in a cohort of 77 adults with spastic diplegia who did not receive SDR. However, because our questionnaire was not validated, the results of this part of the study should be interpreted with caution, and no definitive conclusion can be drawn regarding participation.

### **Gross Motor Function Classification System**

The GMFCS system has been validated as a stable classification system in children (Palisano *et al.*, 2006) and adults (McCormick *et al.*, 2007), and no changes in GMFCS level are normally expected after an intervention (Rosenbaum *et al.*, 2008). However, our data suggest that this system can possibly measure changes in gross motor function over time after dorsal rhizotomy because 64% of our participants (n=10) improved 1 level in the GMFCS, whereas 29% (n=3) remained unchanged and 1 participant (7%) deteriorated from level III to IV 20 years after SDR (Figure 4.4). This is in accordance with a study by Cole *et al.* (2007), who used the GMFCS to show functional improvements in a cohort of 19 patients (17 of whom were diagnosed with diplegia) 18 months after SDR. This cohort (GMFCS level II: n=6, level III: n=10, level IV: n=3) showed an improvement of at least 1 GMFCS level for 79% of their patients, whereas 21% remained at the same level.

Because the primary aim of our study (and the study of Cole *et al.* (2007) was not to determine if the GMFCS is a reliable and valid tool to measure changes in motor function after an intervention, further research is needed to establish whether the GMFCS could in the future play this important and practical role. In addition, the application of the GMFCS levels retrospectively still requires validation.

### **Study Limitations**

Because this 20 year follow-up study was conducted only on a group of patients with spastic diplegia who were operated in 1985, our sample size was relatively small. Therefore, we were unable to account for confounding factors, which could have influenced our outcome measures. One of these possible factors is further orthopaedic surgery after SDR, which is normally performed in 65% of the patients (Steinbok, 2001) and is comparable to our incidence rate of 64%. Therefore, it is impossible to conclude that the improvements we found in neuromuscular outcomes (*Body Function & Structure*) and functional outcomes (*Activity*) are exclusively caused by dorsal rhizotomy. However, we would argue that SDR played a decisive and critical role in achieving these improvements.

A second limitation of the study is the absence of data with which to compare our results. Our study did not include a control group of patients who did not receive

SDR, which would have allowed us to interpret the outcomes from a better perspective. In addition, most studies on adults with CP do not distinguish between the types of CP, which makes it impossible to compare our data with their outcomes. Furthermore, only short-term follow-up studies have been published comparing SDR with other interventions such as orthopaedic surgery<sup>31</sup> or intrathecal baclofen pump implantation (Kan *et al.*, 2008), which limited us in comparing our results with other research groups and their long-term outcomes of different interventions.

A third and final limitation of this study was that we had no freedom in the selection of our assessment tools because the preoperative measurements were conducted in 1985. This is nevertheless inherent to long-term follow-up studies, whereas adapted or new assessment tools are always developed over time. The tests and ordinal assessment scales that we used were developed and described in detail by Berman *et al.* (1989, 1990) (and have been reproduced here in the Appendices) but have never been tested for validity or reliability. The 2 physiotherapists (N.G.L., R.P.L.) who applied the assessment scales 20 years after SDR were nevertheless in agreement and had confidence in their judgments.

## CONCLUSIONS

As presented in our 20 year follow-up study that focused on gait parameters, patients with spastic diplegia also showed improvements in their functional status based on 2 dimensions of the ICF-model, *Body Function & Structure* and *Activity*, 20 years after SDR. Although we could not control for all confounding factors, we argue that most of these improvements were because of SDR. Outcome measures of a third dimension of the ICF-model, *Participation*, were not directly measured but rather indications of active participation status found through a questionnaire. Although the GMFCS was originally developed as a prognostic classification tool, our study suggests that the GMFCS might play an important role in detecting changes over time after an intervention.

We encourage researchers to perform further long-term follow-up studies in patients with different types of CP. This would enable the comparison of a variety of treatment

options in different diagnostic CP groups, which could ultimately assist clinicians, patients, and parents when considering the most effective treatment option.

## ACKNOWLEDGEMENTS

We wish to thank the patient in Figure 4.3 for giving us permission to use his photographs. Funding was provided by Science Foundation Ireland, the South African Medical Research Council and the University of Cape Town. The study protocol was approved by the University of Cape Town's Human Ethics Committee.

### Appendix I Rating categories for outcome measure of ICF-model *Body Function & Structure*

#### Muscle tone (increase)

1 Hypotonia	The limb feels floppy and is unable to resist gravity.
2 Normal	A good balance between agonist and antagonist. Movement feels controlled. No resistance to passive movement.
3 Mild	There is slight resistance to passive stretch and there is a slight decrease in joint mobility. The stretch reflex occurs when the muscle is in a lengthened position.
4 Moderate	There is a greater resistance to passive movement than in 3 above and a greater decrease in mobility. The stretch reflex occurs in the middle of the range.
5 Severe	There is a severe increase in resistance to passive stretch and a severe decrease in mobility. The stretch reflex occurs when the muscle is in a shortened position.

#### Joint stiffness (limitation)

1 Normal	No contracture. The joint can be moved freely and full range is easily attained
2 Mild	There is some stiffness. This can easily be released by passive movement.
3 Moderate	There is a great deal of stiffness but this can be released with difficulty on passive movement. Full range can be attained.
4 Total	Apparent fixed contracture or fixed contracture. Great limitation to passive movement. Full range of movement of the joint cannot be achieved before surgery but may be achieved after surgery.

## Chapter 4

### Voluntary movement (limitation)

1 Normal	Able to initiate and inhibit (i.e. stop and start) a movement 7 to 8 times during one full range of movement. The movement is smooth.
2 Moderate	Able to initiate and inhibit the movement 5 to 6 times. Can achieve full range, but movement is jerky and mass patterns are present.
3 Moderate	Able to initiate and inhibit the movement 3 to 4 times. Can achieve middle or full range, but movement is not smooth.
4 Moderate severe	Able to initiate and inhibit the movement 1 to 2 times. Movement is jerky or overshoots, or there is total movement and patient uses compensations in attempting the movement.
5 Severe	Unable to initiate and inhibit movement. Range is totally limited, or there is a totally abnormal pattern.

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Reprinted with permission (Berman, 1989)

### Appendix II Rating categories for functional outcome measures

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#### Rolling (limitation)

1 Normal	The patient can roll with good separation between the shoulders and the pelvis. In moving from prone to supine, the pelvis leads, and from supine to prone, the shoulders lead. There is good separation between the legs.
2 Mild	The patient can roll with some separation between the shoulders and the pelvis. There is some separation of the legs and some pelvic mobility.
3 Moderate	The patient rolls without separation between the shoulders and the pelvis but with dissociation between the legs, or with some degree of separation between the shoulders and the pelvis but without dissociation between the legs.
4 Moderate severe	The patient rolls <i>en bloc</i> , without separation between the legs and without separation between the shoulders and the pelvis.
5 Severe	The patient cannot roll independently or may roll only with facilitation by the therapist.

#### Side sitting (limitation)

1 Normal	The patient can assume side sitting and sits adequately, i.e. protraction of the weight bearing hip. Lateral weight shift and elongation of the weight bearing side facilitates lateral righting of the trunk on that side. He is able to free his arms from the support surface.
2 Mild	The patient can assume side sitting with inadequate weight shift onto the weight bearing side (i.e. he sits on both buttocks) due to inability to protract the weight bearing side. He lacks mobility at the pelvis. He can maintain this position. He can free his arms.

- 3 Moderate      The patient can assume the position as in 2 above but is unstable and cannot maintain it. He may or may not be able to free his arms.
- 4 Moderate  
severe      The patient can not assume the site sitting position but can maintain it if the therapist provides compression through the shoulder and elbow of the arm on the weight bearing side and assists with weight transference onto the weight bearing side.
- 5 Severe      The patient can be placed in a site sitting position but needs a lot of external support, and he cannot maintain this position.

## Long sitting (limitation)

- 1 Normal      Patient can long-sit independently. No rounding of the back. Good hip flexion and knee extension. He does not require his arms to “hang on.”
- 2 Mild      The patient can long-sit and uses arm support but can let go momentarily. He long-sits with some rounding of the back.
- 3 Moderate      The patient long-sits with some rounding of the back, poor hip flexion and leg extension, and some internal rotation of the legs. He can support himself on the side.
- 4 Moderate  
severe      The patient long-sits but uses forward arm support. There is a lot of back rounding, poor hip flexion and knee extension, and marked internal rotation of the hips.
- 5 Severe      The patient can long-sit only with assistance from the therapist.

## Prone kneeling (limitation)

- 1 Normal      The quadruped position is assumed and maintained without a lordosis and with good arm support. The thighs are held at an angle of 90 degrees to the trunk while the knees are kept at an angle of 90 degrees to the thighs.
- 2 Mild      The patient can assume and maintain a quadruped position but he has a lordosis and some lower leg abduction. He stabilizes his pelvis with active hip flexion. Arm support is good.
- 3 Moderate      The patient can assume the quadruped position. He maintains hip flexion, adduction and internal rotation, and knee flexion. The lower legs are slightly abducted to stabilize the base. The feet are in a mass dorsiflexion pattern or rest on the medial borders. Arm support is fair to good.
- 4 Moderate  
severe      The patient can assume the quadruped position only with difficulty. The hips remain semi-extended and internally rotated, with adduction or abduction. Mass dorsiflexion of feet may be present. Arm support is poor.
- 5 Severe      The patient cannot assume this position and needs assistance to maintain it. The arms collapse as arm support may be poor.

## Chapter 4

### Kneel standing (limitation)

- |                   |  |
|-------------------|--|
| 1 Normal          | The patient can assume and maintain the kneel standing position and maintain a neutral pelvic tilt.  |
| 2 Mild            | The patient can rise against gravity and may or may not have difficulty maintaining this position. He maintains an anterior pelvic tilt, a lordosis, and hip flexion with lower leg abduction. He has great difficulty shifting weight. He can perform an activity in this position. |
| 3 Moderate        | As in 2 above, but the hips are adducted and internally rotated and there is lower leg abduction. He can maintain this position for a few seconds with little support. He is unable to shift weight and has difficulty freeing his arms.   |
| 4 Moderate severe | The patient assumes or is assisted to assume kneel-standing. He has an anterior pelvic tilt, a lordosis, hip adduction, internal rotation, and flexion. He can maintain this position but is unable to free his hands.   |
| 5 Severe          | He cannot assume this position but needs to be placed in the position and given full external support.   |

### Half kneeling (limitation)

- |                   |  |
|-------------------|--|
| 1 Normal          | The patient can assume and maintain a good half kneeling position. He is able to free his arms for an activity.  |
| 2 Mild            | The patient can assume and maintain the half kneeling position but internally rotates the forward leg and flexes the hip of the weight bearing leg. Legs separate easily and the patient can free his arms.  |
| 3 Moderate        | The patient can assume or is helped to assume half kneeling. The legs separate but the patient needs support, i.e., protraction of the pelvis on the weight bearing side and traction of the forward leg. He cannot free his arms or can do so for only a short while. |
| 4 Moderate severe | The patient can assume this position but can separate his legs with difficulty, and he cannot free his hands. If he cannot assume this position and is placed in it, he can only maintain it with difficulty.  |
| 5 Severe          | The patient cannot assume this position. There is great difficulty separating the legs when he is placed in this position. The therapist must maintain full external support and the patient collapses after a few seconds.  |

### Crawling (limitation)

- |          |   |
|----------|---|
| 1 Normal | The patient crawls reciprocally with well-integrated righting and equilibrium reactions, His legs separate with ease. The weight is well distributed and arm support is good. |
| 2 Mild   | The patient crawls with some reciprocation but uses side flexion instead of elongation of the weight bearing side. Arm support is good.                                       |

3 Moderate	As in 2 above, but the legs shoot into extension on the non-weight bearing side or he crawls with a mass flexion pattern with legs widely abducted at the hips and with flexion at the knees. Arm support may vary.
4 Moderate severe	The patient “bunny-hops,” moving both lower extremities together. Arm support may vary.
5 Severe	The patient is unable to assume the four point kneeling position. He “mermaid” crawls by pulling his body forward with his arms while the legs remain in extension. There is very poor dissociation of the legs. Arm support is usually poor.
Standing (limitation)	
1 Normal	The patient is able to assume and maintain a good standing posture.
2 Mild	The patient can assume the standing position but stands with an anterior pelvic tilt. He can shift his weight using side flexion. He stands independently. Feet may or may not be in a valgus position.
3 Moderate	As in 2 above, but he needs to hold on due to lack of stability. The patient has an anterior pelvic tilt, some internal rotation at the hips with slight flexion, and some knee flexion, The feet may be in a valgus position.
4 Moderate severe	Contractures and deformities are developing at the hips, knees, and ankles. The patient can only assume standing for a few seconds. Feet may be in valgus position.
5 Severe	As in 4 above. The patient cannot assume standing. The patient requires full external support to stand. Hips, knees, and ankles may have fixed deformities. Feet may be in a valgus position.
Walking (limitation)	
1 Normal	The patient walks reciprocally. Starting from a stable standing posture, the body is propelled forward in a smoothly coordinated way. The gait cycle consists of a stance and a swing phase. The stance is a 5-phased activity, involving contact, loading response, mid-stance, terminal stance, and pre-swing. The swing is divided into initial mid-, and terminal thirds.
2 Mild	The patient walks with an anterior pelvic tilt but has the necessary break-up of patterns.
3 Moderate	The patient can walk, has the necessary break-up of patterns, but lacks rotation and has poor weight-shift. He uses upper trunk compensations to assist with gait.
4 Moderate severe	The patient has poor break-up patterns and cannot take a few steps independently or has very poor balance.
5 Severe	No reciprocal gait, no break-up of patterns, and no weight shift. The patient needs full external support.

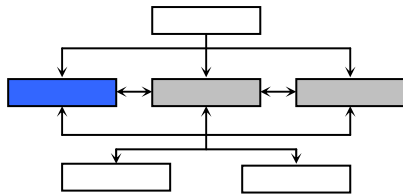
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# Chapter 5



Incidence of spinal abnormalities in patients with spastic diplegia 17 to 26 years after selective dorsal rhizotomy

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## ABSTRACT

**Introduction:** The aim of this study was to evaluate the mechanical status of the spine in patients with spastic diplegia 17 - 26 years after selective dorsal rhizotomy (SDR). **Methods:** We compared original radiographic reports from our earlier short-term follow-up study with current X-rays. In addition, we obtained magnetic resonance images (MRI) of the spine, and additional information regarding back pain and clinical assessments. **Results:** Thirty patients (17 males, 13 females; median age 26.8 years) participated in current study, with median follow-up times of 4.0 and 21.4 years. Comparison of the X-ray results showed respectively: scoliosis 0% and 57%; kyphosis 0% and 7%; lordosis 21% and 40%; spondylolysis 18% and 37%; and spondylolisthesis grade I occurred in 1 patient. The only statistically significant difference was found for scoliosis ( $p < 0.01$ ). The majority had Cobb angles  $< 30^\circ$  with only 2 patients with curves of  $35^\circ$ . MRI-scans showed spinal stenosis in 27%, black discs in 10% and disc protrusion in 3%. Daily back pain was reported in 17%, while 23% reported 'moderate disability' as a result of back and leg pain. No patient to date has required any surgical intervention on the spine. **Conclusions:** Except for spondylolisthesis, spinal deformities did appear to progress with time. However, this increase was not marked, and the development of relatively mild scoliosis was the only statistically significant increase. This group of patients requires continued follow-up. Further studies are required to ascertain the natural history of spinal deformity in adults with spastic diplegia who have not had SDR.

**Keywords:** Cerebral Palsy • Dorsal rhizotomy • Long-term follow-up • Spinal deformity • X-ray • MRI-scan

## INTRODUCTION

The operation of dorsal rhizotomy was first performed in the early 1900's (Foerster, 1913), but gained greater acceptance after Warwick Peacock refined the operative technique in the 1980's, while working at the Red Cross Children's Hospital in Cape Town (Peacock and Eastman, 1981). During the last decade various research groups around the world have shown that selective dorsal rhizotomy (SDR) in patients with cerebral palsy (CP) leads to functional improvements during the early years after surgery (Steinbok, 2007; Farmer and Sabbagh, 2007). Recently we were able to show sustained long term benefits in two 20 year follow-up studies (Langerak *et al.*, 2008; Langerak *et al.*, 2009).

Despite the fact that SDR is invariably successful in its aim of reducing spasticity, spinal instability as a result of the laminectomy remain a concern (Aiona and Sussman, 2004; Steinbok, 2007; Langerak *et al.*, 2008). In the early 1990's we published studies providing incidences of spinal deformities in children with CP after SDR (Peter *et al.*, 1993; Peter *et al.*, 1990). These and 5 other peer-reviewed short-term follow-up studies suggest that the incidence of spinal deformities after SDR varies from 40 - 88%. Deformities include scoliosis in 16-55%, thoracic hyperkyphosis in 1-41%, lumbar hyperlordosis in 7 - 50%, spondylolysis in 9 - 12% and spondylolisthesis in 2 - 24% of the patient cohorts (Peter *et al.*, 1990; Johnson *et al.*, 2004; Steinbok *et al.*, 2005; Turi and Kalen, 2000; Spiegel *et al.*, 2004; Golan *et al.*, 2007; Peter *et al.*, 1993).

However, spinal deformity may also occur as part of the natural history of CP. Morrell *et al.* (2002) reported that patients with CP who did *not* undergo SDR showed an increased incidence of scoliosis with age, accompanied by increased thoracic and lumbar curves in the sagittal plane and the development of spondylolysis and spondylolisthesis. Three other studies based on institutionalized patients with CP also show increased scoliosis with age (Thometz and Simon, 1988; Saito *et al.*, 1998; Majd *et al.*, 1997), while Harada *et al.* (1993) found progressive lordosis and spondylolysis in adults with spastic diplegia. Nevertheless, we do not know whether patients who underwent SDR in childhood have a greater risk for developing spinal

deformities over time, as there are no long term studies that report these changes in adulthood.

Therefore, we aimed to examine the results of clinical, radiographic and magnetic resonance imaging (MRI) examinations of patients with spastic diplegia more than 15 years after SDR to determine the long term mechanical status of the spine. In addition, we obtained information regarding the SDR procedure, patients' demographic characteristics, back pain and outcomes of clinical assessments to further interpret the results of the radiological studies.

## **METHODS**

### ***Patients***

We performed a retrospective review of all patients who underwent SDR between 1981 and 1991 at Red Cross Children's Hospital in Cape Town. In order to facilitate follow-up we restricted the cohort to those who lived within a radius of 100 kilometers of Cape Town at the time that the operation was performed. Based on detailed inclusion and exclusion criteria (Table 5.1) a neurosurgeon (JCP) and physiotherapist (NGL) completed the first patient selection. There were 106 patients operated on between 1981 and 1991 of whom 47 met the inclusion criteria. With help from other clinicians, patients, patients' families, teachers and physiotherapists at schools and an internet search, we were able to track down 37 of these 47 patients (79%). Thirty patients (64%) agreed to participate in the current study, while 7 (15%) chose not to participate (for reasons other than health status).

This study was approved by the Human Ethics Committee at the University of Cape Town, and we obtained informed consent from each participant.

### ***Study Design***

Data collection took place in 2008 in five different stages and included two visits by the patients: (i) NGL reviewed the historical radiographic reports, which were part of our former short-term follow-up studies (Peter *et al.*, 1993; Peter *et al.*, 1990); (ii) X-rays and MRI-scans of the spine were performed at the Medi-Clinic in Stellenbosch;

**Table 5.1** Inclusion and exclusion criteria for selection of study cohort.

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Inclusion criteria
1. Diagnosis: CP
1.1. Symptom type: spastic
1.2. Limb distribution: mainly involvement of lower limbs or with only mild involvement of
2. Ambulant before age of 4 years (with or without walking aids and/or support)
3. SDR between 1981 and 1991 in Cape Town
4. Goal of SDR operation
4.1. Functional improvement
4.2. Improving gait pattern or, if child was younger than 4 years and not ambulant pre-
5. Attended a school in Cape Town
6. Preoperative physiotherapeutic assessments form available
7. Currently living in Cape Town or neighbourhood (max 100km)
Exclusion criteria
1. Diagnosis: cerebral palsy
1.1 Symptom type: dystonic, athetotic, ataxia, and hypotonic
1.2.Total body involvement
2. Diagnosed with other neuromuscular disorders

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(iii) a neurosurgeon (JCP) and an orthopaedic surgeon (EBH) examined the scans, blinded to the results of the previous imaging, and consulted two other neurosurgeons (AGF and AAF) if there was any doubt; (iv) clinical assessments were performed by a neurosurgeon (JCP) and physiotherapist (NGL); and (v) patients completed the Oswestry Disability Index (ODI) (Fairbank *et al.*, 1980) and a self-developed questionnaire which focused on the frequency of spinal, upper and lower extremity pain.

### **Outcome Measures**

#### *X-rays and MRI-scans*

The method of examination and evaluation of the radiographs was similar to our short-term studies (Peter *et al.*, 1993; Peter *et al.*, 1990). X-rays were taken in a standing position (or sitting erect if not possible to stand in the first study) holding onto a bar if necessary with antero-posterior (AP), lateral and oblique views. In the frontal plane scoliosis was measured by Cobb angles (Cobb, 1948). X-rays in the

sagittal plane were used to evaluate thoracic kyphosis (T3-T12) and lumbar lordosis (L1-L5). In line with our previous study (Peter *et al.*, 1990), the ranges described by Propst-Proctor and Bleck (1983) were used as a reference for normal; kyphosis 12° - 40° and lordosis 23° - 54°. The incidence of spondylolysis and spondylolisthesis was examined by using lateral and oblique radiographic views. Spondylolisthesis was measured as a percentage of displacement, and graded by the Meyerding system (Meyerding, 1932): grade I (25%), II (50%), III (75%) and IV (100%).

The lumbar spines were examined with MRI (Siemens Magnetom Symphony 1.5 Tesla, Germany) and a body coil. The imaging protocol consisted of the following: (i) sagittal T1 and T2 weighted fast spin-echo from mid-thoracic to sacrum; (ii) axial T2-weighted fast spin-echo from L1-S2; and (iii) coronal T1-weighted fast spin-echo from L1-S2. MRI-scans were examined for spinal stenosis, black discs, and disc herniation. In addition to these degenerative disorders, the level of the conus and incidence of any other spinal abnormality were evaluated (Modic, 1994).

### *Clinical Assessments*

Clinical examination included evaluation of muscle tone, muscle strength, skin sensation and deep tendon reflexes in both lower extremities (Table 5.2). Muscle tone was assessed using the Ashworth Scale (Ashworth, 1964). Muscle strength assessments were scored using the MRC scale (Medical Research Council, 1943). Sensation was tested for touch, pin-prick and deep pressure corresponding to dermatomes from L1 to S4/5. Proprioception was recorded as normal or abnormal. The reflexes evaluated included the plantar reflex and deep tendon reflexes of the hip adductor, rectus femoris, patellar and achilles tendons. Scores for the plantar reflex were recorded as either positive or negative, while the outcomes of the deep tendon reflexes were classified by values from 0 - 4.

In addition, patients' demographics and general clinical characteristics were evaluated. CP diagnosis was confirmed and body weight and height were measured, which resulted in their Body Mass Index (BMI) (underweight: <18.5; normal: 18.5 - 24.9; overweight 25.0 - 29.9; obese: >30.0) (World Health Organisation Expert Committee, 1995). Patients were also questioned about their walking ability, physical problems related to possible sensory loss (bowel, bladder, sexual problems), and if

they had received any other orthopaedic interventions. Patients were also classified according to the Gross Motor Functional Mobility Scale (GMFCS) (Palisano *et al.*, 1997; Jahnsen *et al.*, 2006), based on the assessors' observation and self evaluation by the patient.

**Table 5.2** Description and scores for clinical assessment tools.

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Spasticity: Original Ashworth scale	
0	No increase in tone
1	Slight increase in tone giving a catch when the limb is moved in flexion or extension
2	More marked increase in tone but limb easily flexed
3	Considerable increase in tone, passive movement difficult
4	Limb rigid in flexion or extension
Muscle strength: MRC scale	
0	None
1	A palpable contraction
2	Motion with gravity eliminated
3	Antigravity strength
4	Some resistance present
5	Normal strength
Sensory abnormalities	
0	None
1	Loss of pin-prick sensation (dermatomal)
2	Patchy irregular diminution of sensation (non-dermatomal).
Deep tendon reflexes	
0	Absent despite reinforcement
1	Present only with reinforcement
2	Normal
3	Increased but normal
4	Markedly hyperactive,with clonus

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### *Pain Questionnaires*

We used the ODI version 2.0 which is a valid and reliable condition-specific outcome measure for back or leg pain (Fairbank *et al.*, 1980; Fairbank and Pynsent, 2000). The questionnaire includes 10 sections regarding pain, personal care, lifting, walking,

sitting, standing, sleeping, sex life, social life and traveling. The patients were asked per section to choose one statement which most applied to them, ranging from no problems (score 0) to severe problems (score 5), or they had to indicate that a section was not applicable. When the questionnaire was completed a percentage was calculated and patients were classified based on their back pain problems during daily activities into: 0 - 20% indicating 'minimal disability', 20 - 40% 'moderate disability', 40 - 60% 'severe disability', 60–80% 'housebound', and 80 - 100% 'bed bound (Fairbank *et al.*, 1980).

We also developed a short questionnaire which focused on the frequency of pain in different parts of the body. Questions were related to pain at spinal level and upper and lower extremities. Frequency of pain per body part could be scored as: never; occasionally; once per month; once per week; daily; or varies (occasionally to several times per week).

### **Statistical analysis**

Data were analyzed using Stata Version 10.0 (Stata Corporation, College Station, USA). We used nonparametric statistics because our sample size was small and not normally distributed. The Wilcoxon signed-rank test was performed to explore changes over time (short-term versus long-term) for the X-ray outcome measures (incidence of scoliosis, kyphosis, lordosis, spondylolysis and spondylolisthesis). A Fisher exact test was used to examine which factors were associated with the radiographic outcomes. Significance levels were set to  $p \leq 0.05$ .

## **RESULTS**

### **Patients**

The study cohort consisted of 30 patients with spastic diplegia (5 patients had mild unilateral arm involvement) of whom 17 were male and 13 were female. Seventy percent of patients had laminectomies performed from L1/2 to S1, 10% at levels L1-L5, 10% at L2-L5, 3% at L2-S2 and 7% at L3-S1. The median age of the study cohort at time of SDR was 5.2 years +/- 5 years (range 2 - 27 years), and during the current

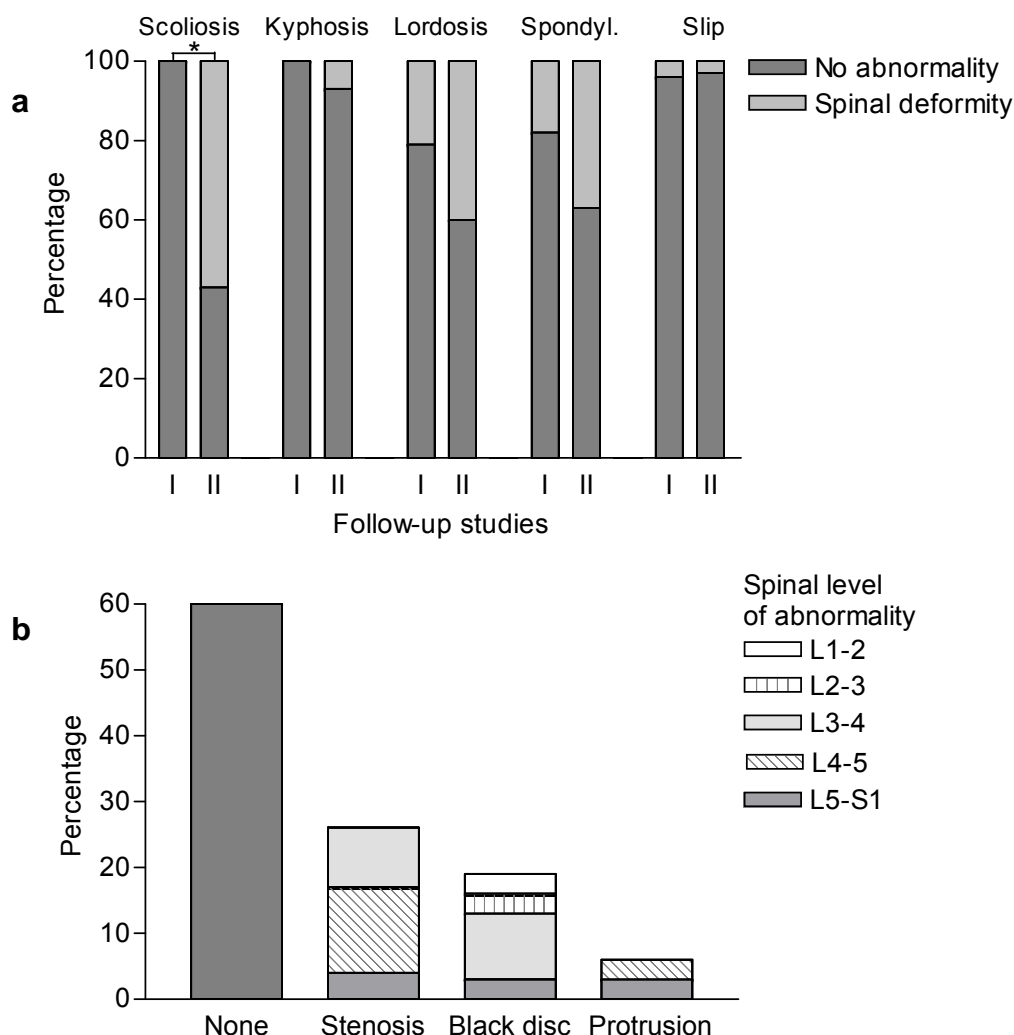
study they were aged 26.8 years  $\pm$  5 years (21 - 44 years). The median short-term follow-up time in this cohort of the original study was 4.0 years  $\pm$  2 years (1 - 8 years), while the mean current (long-term) follow-up time was 21.4 years  $\pm$  3 years (17 - 26 years). All patients were ambulant at follow-up, and the walking ability varied: 20 patients (67%) walked without walking devices, 4 patients (13%) used 1 or 2 crutches only outdoors, and 6 (20%) patients always had to use their crutches. Half were classified as GMFCS level I, 9 patients (30%) as level II and 6 patients (20%) as level III.

The mean BMI of the study cohort ranged between 18 and 36; 2 patients (7%) were classified as underweight, 16 (53%) as normal, 6 (20%) as overweight and 6 (20%) as obese. None of the patients reported problems with sexual, bowel or bladder function. One patient was diagnosed with Crohns' disease and another patient with Graves' disease. This latter patient required oral baclofen medication regularly to reduce muscle spasms. None of the patients who participated in this study ever received intrathecal baclofen or botox before or after SDR. Before SDR, at least 1 orthopaedic surgical operation had been performed on almost half of the cohort (48%), which included muscle releases in 14 patients (48%) and osteotomies of the femur or foot/toes in 3 patients (10%). After SDR, 18 patients (62%) received orthopaedic interventions including muscle releases and osteotomies in 59% and 31% of the study cohort, respectively.

### ***X-rays and MRI-scans***

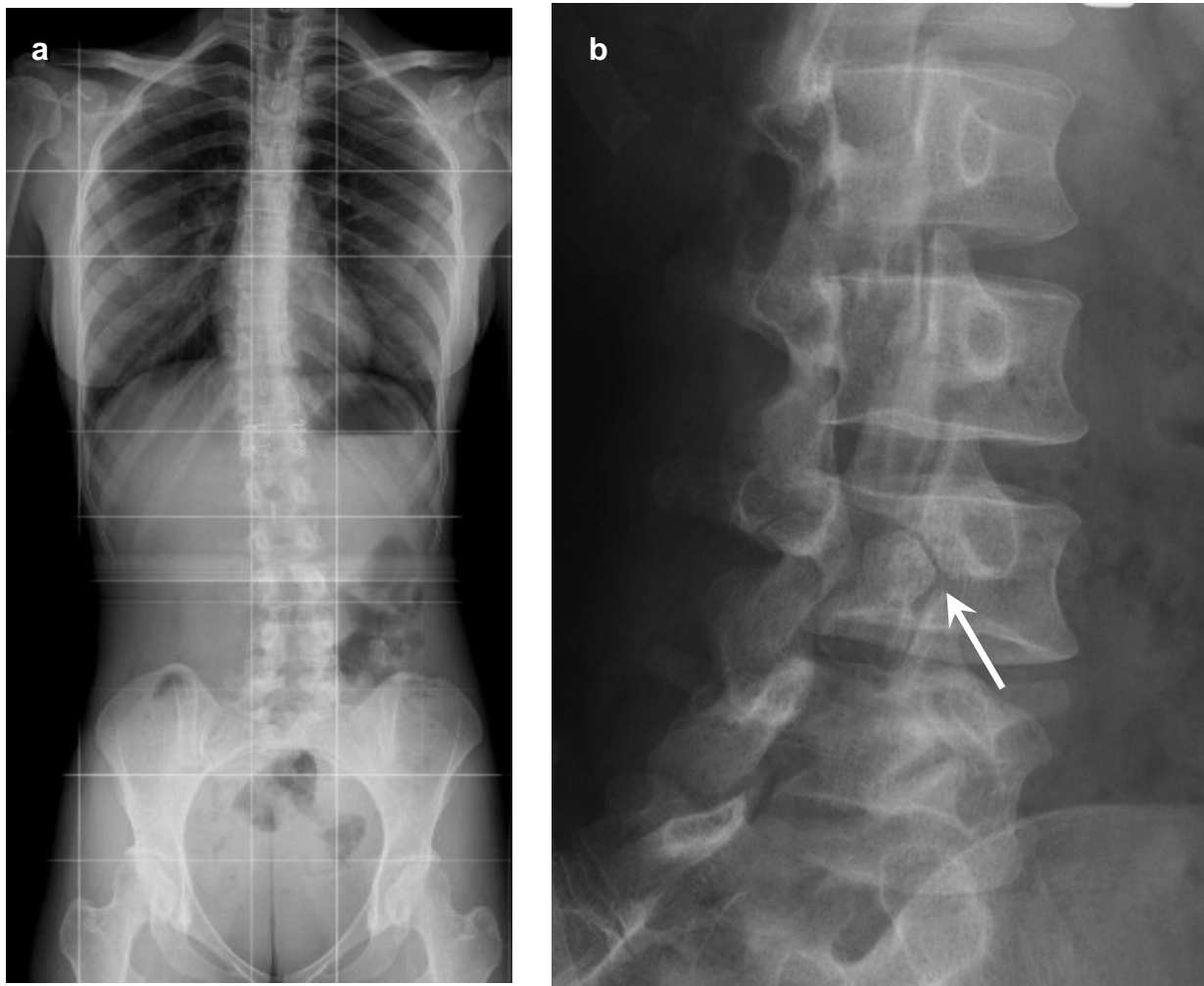
Two of the 30 patients did not take part in our short-term follow-up studies (Peter *et al.*, 1993; Peter *et al.*, 1990) and so historical comparison of imaging was only available for 28 patients. Of these 28 patients, none had been initially diagnosed with scoliosis, while at the long-term follow-up study 15 of the 30 patients (50%) had a scoliosis curve of  $<30^\circ$  and 2 (7%) had curves of  $35^\circ$  on the AP X-rays (Figures 5.1a and 5.2a). Scoliosis was significantly more common in the long term follow-up study ( $p < 0.01$ ). During our initial study none of the patients were diagnosed with kyphosis and 6 patients had lordosis, while in the present study kyphosis and lordosis occurred in 2 (7%) and 12 (40%) patients, respectively. Although thoracic kyphosis and lumbar lordosis were more frequent at follow-up, these differences were not significant ( $p = 0.32$  and  $p = 0.13$ , respectively). Spondylolysis was diagnosed in 5

patients (11% at L3-4; and 7% at L5-S1) at the short-term follow-up. Despite the fact that 4 of these patients showed healing of their pars interarticularis, in the long-term spondylolysis was reported in 11 patients (15% at L2-3; 38% at L3-4; 31% at L4-5; and 13% at L5-S1), which was not significantly different from the initial study ( $p=0.13$ ) (Figures 5.1a and 5.2b). At the first study 1 of the 5 patients had spondylolysis bilaterally at the same level, while in the present study 5 of the 11 patients with spondylolysis were bilateral. Spondylolisthesis Grade I was found in both the short- and long-term follow-up study in only 1 patient, who was also diagnosed with bilateral spondylolysis at level L3-4 during the first and last study (Figure 5.1a).

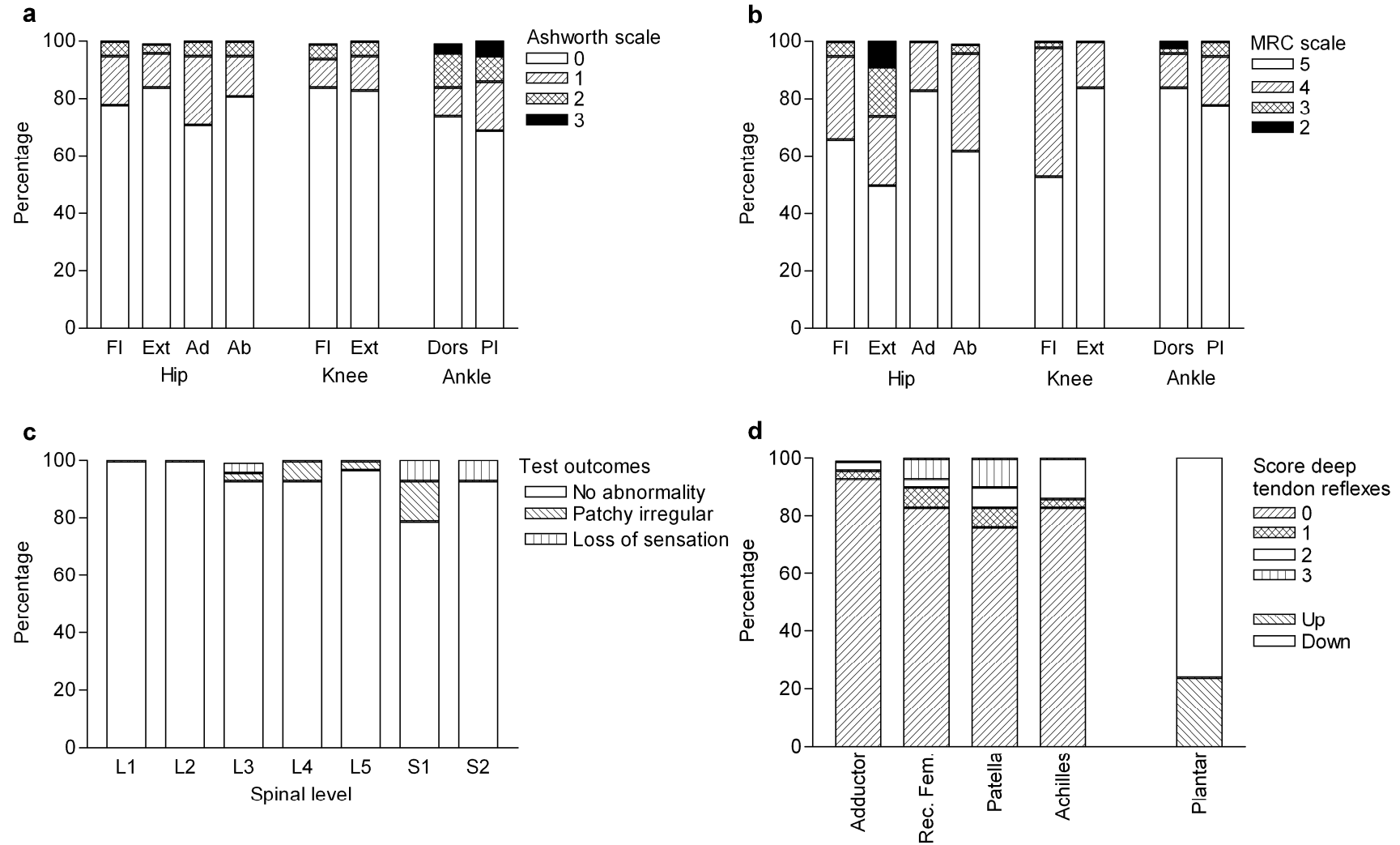


**Figure 5.1** (a): Outcomes of X-rays at short-term (I) (n=28) and long-term (II) (n=30) follow-up studies; and (b): outcomes of MRI-scans at long-term follow-up study. Note: Incidences of scoliosis includes relatively mild scoliosis with the majority of curves  $<30^\circ$  and 2 curves  $35^\circ$ . Spondyl. is an abbreviation of spondylolysis and slip is corresponding to Grade I spondylolisthesis. \*Means that there was a significant difference with  $p<0.05$ .

As shown in Figure 5.1b, 18 of the 30 patients (60%) who participated in the long-term follow-up study did not have any abnormalities on their MRI-scans. Lumbar stenosis was seen in 8 patients (27%) (involving 2 levels in 2 patients, and 3 levels in 1 patient), black discs were seen in 6 patients (20%), and 2 patients had disc protrusions (7%), but no disc extrusion or sequestration. Two patients had combined lumbar stenosis and black discs, and all three abnormalities were seen in 1 patient (without any spinal deformity on X-ray). No other abnormalities were found. The conus was situated at T12 in 2 patients, at L1 in 18, at the lower border of L1 in 1, and at L2 in 9 patients. None of the patients who participated in the current study required any spinal operations after their SDR.



**Figure 5.2** a: AP X-ray of a typical patient with mild scoliosis; and b: an oblique X-ray of a patient with spondylolysis.



**Figure 5.3** Outcomes of the clinical assessments (n=29): a muscle tone; b muscle strength; c pin-prick-sensation; and d deep tendon reflexes. Abbreviations: FI: flexion; Ext: extension; Ad: adduction; Ab: abduction; Dors: dorsi flexion; and Plant: plantar flexion.

### **Clinical Assessments**

One patient was not available for the second assessment, and so we only had clinical assessment outcomes of 29 patients. As is apparent from Figure 5.3a and 5.3b, the majority of our study cohort had normal tone (Ashworth scale 0) and strength (MRC 5) in the various muscle groups evaluated. No patients had abnormalities in proprioception, touch or deep pressure, and most patients had no impaired pin-prick sensations (Figure 5.3c). In addition, most patients had reduced deep tendon reflexes and positive plantar reflexes (Figure 5.3d).

### **Pain Questionnaires**

All 30 patients who participated in the long-term follow-up study completed the ODI. Twenty-three patients (77%) scored 0 - 20% ('minimal disability' because of back pain), while the other 7 patients scored 20 - 40% ('moderate disability'). The outcomes of our self-developed questionnaire are presented in Table 5.3. Few patients (3 - 10%) reported pain in their upper extremities, but 23 - 40% of the patients had pain in their lower extremities. None of the patients suffered from pain in the upper thorax, while 20% of the cohort reported pain at cervical and 67% at lumbosacral spinal level. Pain in the lower back (lumbosacral region) was most common in our study cohort and was reported as occurring on a weekly basis in 2 patients (7%) and daily in 5 patients (17%).

**Table 5.3** Frequency of pain distributed over different parts of the body.

<i>Location of pain</i>	<i>Never</i>		<i>Occasionally</i>		<i>Monthly</i>		<i>Weekly</i>		<i>Daily</i>		<i>Varies<sup>φ</sup></i>	
	<i>n</i>	<i>%</i>	<i>n</i>	<i>%</i>	<i>n</i>	<i>%</i>	<i>n</i>	<i>%</i>	<i>n</i>	<i>%</i>	<i>n</i>	<i>%</i>
<b>Spinal level</b>												
Cervical	24	80	3	10	0	0	1	3	1	3	1	3
Thoracic	30	100	0	0	0	0	0	0	0	0	0	0
Lumbosacral	10	33	10	33	2	7	2	7	5	17	1	3
<b>Upper extremity</b>												
Arm	29	97	0	0	0	0	0	0	0	0	1	3
Shoulder	27	90	1	3	1	3	0	0	1	3	0	0
<b>Lower extremity</b>												
Leg	18	60	10	33	0	0	0	0	0	0	2	7
Hip	26	87	3	10	0	0	0	0	0	0	1	3
Knee	20	67	9	30	0	0	0	0	0	0	1	3

Varies<sup>φ</sup> means with a frequency of pain between occasionally and several times a week.

Pain was reported on a weekly or daily basis in 7 of the 17 patients with scoliosis (41%), 1 of the 2 with kyphosis, 4 of the 12 with lordosis (33%), 3 of the 11 with spondylolysis (27%) and in the patient who had spondylolisthesis. In addition, daily back pain was reported by 1 of the 8 patients with spinal stenosis, 1 of the 6 with a black disk and 1 of the 2 patients with disc protrusion. Eight patients of the current study cohort used medication intermittently to relieve their pain.

### ***Associations with Spinal Deformities***

Table 5.4 shows the frequency of scoliosis, lordosis and spondylolysis for each different variable related to the SDR operation, patients' demographic characteristics, MRI-scan outcomes and pain questionnaire. Since kyphosis (n=2) and spondylolisthesis (n=1) were rare, we did not include these conditions in our statistical analysis. Scoliosis, lordosis and spondylolysis were more common when SDR was performed at a young age (2 - 5 years) compared to older age (>13 years). These differences were smaller between age groups 2 - 5 years and 6 - 12 years for scoliosis and spondylolysis, while the incidence of lordosis was still twice as high in the youngest group. In addition, patients who were currently aged between 21 and 25 years had a relatively higher incidence of scoliosis and spondylolysis compared to patients in the oldest age group (>31 years old), while lordosis was diagnosed mostly in the age group 26 - 30 years old. However, there was no statistically significant difference in the frequency of spinal deformities for the 2 different follow-up times (16 - 20 years and 21 - 26 years).

The 5 patients diagnosed with mild unilateral involvement of an upper extremity were significantly more likely to have spondylolysis than patients with pure spastic diplegia ( $p=0.05$ ), but no major differences were seen for scoliosis and lordosis. Females and males had about the same incidence of scoliosis and spondylolysis, but almost twice as many females compared with males were diagnosed with lordosis. Patients' level of BMI and functioning (GMFCS) was not significantly related to the spinal deformities. However, patients with a higher BMI had a slightly lower frequency of spondylolysis, and spondylolysis was more common in patients with higher levels of functioning (GMFCS I and II).

Tale 5.4 Associations.

	Variables (n)	Scoliosis		Lordosis		Spondylo- lysis	
		%	p	%	p	%	p
<b>SDR</b>							
Age at SDR	2 - 5yr (17)	65	0.68	53	0.27	41	0.87
	6 - 12yr (8)	50		25		38	
	>13 yr (5)	40		20		20	
Follow-up time	16 - 20yr (12)	58	1.00	33	0.71	42	0.71
	21 - 26 yr (18)	56		44		33	
<b>Demographic characteristics</b>							
Current age	21 - 25 (10)	60	0.22	30	0.15	50	0.51
	26 - 30 (11)	73		64		36	
	>31 (9)	33		22		22	
CP with unilateral Upper limb involvement	No (25) Mild (5)	56 60	1.00	40 40	1.00	28 80	0.05*
Gender	Male (17)	59	1.00	29	0.26	41	0.71
	Female (13)	54		54		31	
BMI	<18.5 (2)	100	0.59	50	0.84	50	0.47
	18.5 - 24.9 (14)	57		43		50	
	25.0 - 30.0 (8)	38		25		25	
	>30.0 (6)	67		50		17	
GMFCS	I (15)	53	0.79	33	0.70	40	0.69
	II (9)	67		44		44	
	III (6)	50		50		17	
<b>MRI-scan outcomes</b>							
Stenosis (St)	No (22)	55	1.00	41	1.00	27	0.10
	Yes (8)	63		38		63	
Black disc (Bd)	No (24)	54	0.67	42	1.00	33	0.64
	Yes (6)	67		33		50	
Disc protrusion (Dp)	No (28)	57	1.00	43	0.50	36	1.00
	Yes (2)	50		0		50	
Number of abnormal- ities per patient	No abnormalities (18)	50	0.46	44	0.91	22	0.04*
	1: St or Bd or Dp (9)	67		33		56	
	2: St and Bd / Bd and Dp (2)	100		50		100	
	3: St and Bd and Dp (1)	0		0		0	
<b>Pain questionnaire</b>							
ODI	0 - 20 (23)	52	0.43	39	1.00	30	0.37
	21 - 40 (7)	71		43		57	

Incidences (%) and associations (p-values) of each variable for the different spinal deformities as diagnosed at the long-term follow-up study were calculated; Scoliosis (n=17), Lordosis (n=12) and Spondylolysis (n=11). \*Significant differences in incidences of certain spinal deformity between the variables as tabulated (Fisher exact test:  $p < 0.05$ ).

Disc abnormalities were not associated with radiographic spinal abnormalities. The overall number of spinal abnormalities found with the MRI-scans only showed a marginally significant relationship with spondylolysis ( $p=0.04$ ). No significant relationships were seen between the outcomes of the validated pain questionnaire and spinal deformities.

## DISCUSSION

This follow-up study provides information about the incidence of spinal abnormalities based on X-rays and MRI-scans in patients with spastic diplegia (or with minor unilateral involvement of an upper extremity) more than 15 years after they received SDR. Extending the results of short-term follow-up studies after SDR (Peter *et al.*, 1990; Johnson *et al.*, 2004; Steinbok *et al.*, 2005; Turi and Kalen, 2000; Spiegel *et al.*, 2004; Golan *et al.*, 2007), we report further progression of spinal deformities (scoliosis, kyphosis, lordosis, spondylolysis) in the long-term. However, only the incidence of relatively mild scoliosis was statistically significantly increased, and no patients with spondylolysis developed spondylolisthesis. In 60% of our study cohort we found no MRI abnormalities. The total number of disc degeneration and spinal stenosis was statistically significantly associated with spondylolysis, as well as the diagnosis of spastic diplegia with mild unilateral upper limb involvement. Seventy-seven percent of the study cohort had no or minimal disability because of back or leg pain, and none of the patients required any other spinal interventions.

### **Scoliosis**

Studies of CP patients report various ranges for the incidence of scoliosis, from 38% of patients with CP aged between 14 and 19 (Rosenthal *et al.*, 1974), 54% in patients older than 20 years (Robson, 1968), to 64% for institutionalized adults (Madigan and Wallace, 1981). Short-term follow-up studies after SDR (mean follow-up time 4-8 years) describe scoliosis in 17 - 55% of patients based on sitting and standing radiographs (Johnson *et al.*, 2004; Steinbok *et al.*, 2005; Golan *et al.*, 2007; Spiegel *et al.*, 2004; Turi and Kalen, 2000; Peter *et al.*, 1990).

None of the patients who also participated in our original follow-up study had scoliosis at 4 years (range 1 - 8 years) after SDR. In the current study we found an incidence of scoliosis of 57% at 21.4 years (range 17 - 26 years) after SDR. This increase was statistically significant between the short- and long-term follow-up. However, the majority had curves  $<30^\circ$  and only 2 had curves of  $35^\circ$ . Steinbok *et al.* (2005) (based on standing X-rays) also found the incidence of scoliosis to be 55% and 6% of their patients had curves  $>35^\circ$  in a short-term study.

Some studies based on institutionalized patients with CP have reported progression of the scoliotic curve with age (Saito *et al.*, 1998; Thometz and Simon, 1988; Majd *et al.*, 1997; Thometz and Simon, 1988). In our study however, the oldest patients had the lowest incidence of scoliosis, and there was no increased incidence with longer follow-up time. However, younger age at SDR (2 - 5 years old) had a higher incidence of scoliosis. This is in line with the results of other short-term data (Steinbok *et al.*, 2005). On the other hand, this concern might be balanced by the need for less lower limb orthopaedic surgery in younger patients (O'Brien and Park, 2006).

### **Kyphosis**

As far as we could discern, there is very little published on the natural history of kyphosis in adults with CP. Most short-term follow-up studies after SDR described hyperkyphotic curves between 1 - 16% (Turi and Kalen, 2000; Spiegel *et al.*, 2004; Johnson *et al.*, 2004; Golan *et al.*, 2007; Peter *et al.*, 1990), while one study reported kyphosis in 41% of the patients (Steinbok *et al.*, 2005).

None of our patients were diagnosed with kyphosis in the initial short-term follow-up study. However, overall in the long-term 7% had kyphosis defined as a thoracic curve  $>40^\circ$ . Compared to our own and other short-term follow-up studies, this suggests that there is minimal progression of kyphosis with time. Since only 2 patients in this study were diagnosed with kyphosis we were not able to determine associations between different variables.

### **Lordosis**

Harada et al. (1993) found in their 84 patients with spastic diplegia an average curve for lumbar lordosis of 54°, which is considered to be the upper limit for normal. In short-term follow-up studies after SDR, the incidence of hyperlordosis varied from 7 - 50% using standing X-rays (Spiegel *et al.*, 2004; Johnson *et al.*, 2004; Golan *et al.*, 2007; Steinbok *et al.*, 2005; Peter *et al.*, 1990), while a combination of standing and sitting (if standing was not possible) X-rays resulted in an incidence of 21% (Steinbok *et al.*, 2005), and another study, where the X-ray position was not described in detail, found 15% (Turi and Kalen, 2000).

An incidence of 21% was found in the patients who participated in our initial short-term follow-up study, while now more than 15 years after SDR we found an overall incidence of 40%. This is higher compared to other short-term follow-up studies after SDR, with the exception of the study published by Johnson et al. (2004), who reported an incidence of 50% after the longest mean follow-up time of 8.6 years.

One of the reasons for the increase in incidence could be the natural history of CP, as Harada et al. (1993) and Steinbok et al. (2005) found a correlation between hyperlordosis and age of the patients. Golan et al. (2007) reported an increased lordosis of 3.6° for each year after SDR, and Johnson et al. (2004) reported a higher incidence with longer follow-up time. Although none of our associations were statistically significant, it appeared that young age at SDR (2 - 5 year) had the highest incidence of lordosis, but patients who were classified in the longest follow-up time group had only an 11% higher incidence. As with scoliosis, these observations need further investigation as other factors (such as the need for orthopaedic surgeries of the lower extremities (O'Brien and Park, 2006)) should also be taken into account before age criteria for SDR are changed. Like Golan et al. (2007), we also found a higher incidence of lordosis in females, although this was not statistically significant.

### **Spondylolysis and Spondylolisthesis**

Frederickson et al. (1984) reported spondylolysis in 4% of the children in a normal population, which increased to 6% in adulthood, and 74% of these patients also showed spondylolisthesis. These results were however based on X-rays taken in a

supine position which underestimates the true incidence (Fredrickson *et al.*, 1984). Oblique and lateral standing radiographs are necessary to detect the true incidence (Lowe *et al.*, 1976). Harada *et al.* (1993) reported (using standing X-rays and oblique views) spondylolysis at level L5 in 21% of a cohort of patients with spastic diplegia, in which two-thirds were older than 20 years. They also found spondylolisthesis in 4% compared to 6% in the healthy control group. The short-term SDR follow-up studies, where no oblique views were used, reported (mainly at level L5) spondylolysis in 12% (Golan *et al.*, 2007), spondylolisthesis in 2 - 24% (Turi and Kalen, 2000; Spiegel *et al.*, 2004; Johnson *et al.*, 2004; Golan *et al.*, 2007). In our post SDR follow-up studies we always included standing X-rays with oblique views and the first study showed spondylolysis in 9% and spondylolisthesis in 4% (Peter *et al.*, 1990), while at follow-up these were 13% and 6% respectively (Peter *et al.*, 1993).

The patients who were eligible for our current follow-up study progressed from 18% in the short-term to 37% in the long-term. Spondylolisthesis grade I was diagnosed in the same patient as in the short- and long-term follow-up study with no progression. Spondylolysis was more common in this long-term study than in the other short-term studies. We found these abnormalities at different levels between L2-S1, while other short-term follow-up studies reported abnormalities mainly at L5-S1. When compared with X-ray results of spastic diplegia patients who did not undergo SDR (using similar radiographic methods) (Harada *et al.*, 1993), spondylolysis was slightly more common in our study, but the patients may not be exactly comparable. Despite the fact that 4 patients showed spontaneous healing of the defect in the pars interarticularis, as also reported by other research groups (Wiltse *et al.*, 1975).

The development of spondylolysis is in most cases a result of a fatigue fracture of the pars. Patients who are not ambulant have a much lower chance of developing spondylolysis and spondylolisthesis than the more functionally mobile patients. This is supported by the results of Rosenberg *et al.* (1981), who reported that none of their non-ambulant patients developed spondylolysis or spondylolisthesis. In line with this, spondylolysis was less common in our study in patients who were classified as GMFCS level III. In addition, we found that patients with mild involvement of an upper limb had a significantly higher incidence of spondylolysis than the patients with no upper limb involvement, which might be related to more asymmetrical strain

experienced by the lumbar spine. With regard to the MRI outcomes, spondylosis was more common in patients who had spinal stenosis (not significant), compared to those without this spinal abnormality. In addition, a statistically significant association was found for the total number of MRI abnormalities and spondylolysis.

### ***MRI-scan Outcomes***

Degenerative changes of the spine in adults with CP have been described in the literature (Harada *et al.*, 1993; Rosenberg *et al.*, 1981), although there is an incomplete overview of the incidence of spinal abnormalities other than the bony spinal deformities.

Disc degeneration is a result of progressive structural failure, which is often the result of aging or excessive loading of the spine (Adams and Roughley, 2006). Asymmetrical and increased strain and excessive mobility are all described in the pathogenesis of spinal stenosis and most of our cases with spinal stenosis appear to have been caused by facet hypertrophy. In addition, we found that spinal stenosis was most common at level L4-5 but sometimes occurred at multiple levels, which is in accordance with what is described in the literature (Modic, 1994).

### ***Clinical Assessments***

Different short-term (Farmer and Sabbagh, 2007; Steinbok, 2007) and long-term (Langerak *et al.*, 2009) follow-up studies provided convincing evidence that muscle tone is significantly decreased and that muscle strength might deteriorate, remain unchanged, or improve as a result of SDR. Persistent sensory loss has been reported in 23% of children with CP (McLaughlin *et al.*, 2005), and in 4 - 20% in short-term SDR follow-up studies (Steinbok and Schrag, 1998; Peter and Arens, 1993). In addition, significant decreased or absent deep tendon reflexes have been reported in most patients in short-term follow-up studies (Nordmark *et al.*, 2008). The outcomes of muscle tone, strength, sensation and reflexes of our current long-term study were similar.

### ***Pain Questionnaires***

Low back pain and pain in the lower extremities were the most commonly reported sites for pain in adults with CP (Jahnsen *et al.*, 2004; Opheim *et al.*, 2009; Turk *et al.*,

1997; Schwartz *et al.*, 1999; Engel *et al.*, 2003; Hilberink *et al.*, 2007; Andersson and Mattsson, 2001). In a mixed study, cohorts of patients with CP aged from 18 - 70+ years, back pain was reported in 54 - 67% of the cases (Jahnsen *et al.*, 2004; Opheim *et al.*, 2009; Schwartz *et al.*, 1999; Engel *et al.*, 2003). Some studies found that this pain had no or minor impact on patients' daily functioning (Hilberink *et al.*, 2007; Schwartz *et al.*, 1999; Engel *et al.*, 2003), while another study showed that this had a moderate to extreme impact on one-third of their patients' activities (Jahnsen *et al.*, 2004). These outcomes are similar to our results of 67% of our adults who reported back pain (17% on a daily basis), and 23% of them who were 'moderately' disabled because of back or leg pain more than 15 years after SDR.

In addition to these more descriptive articles, back pain has also been discussed in radiographic spinal deformity studies. Short term SDR follow-up studies in children found back pain in 0 - 29% of the patients (Steinbok and Schrag, 1998; Spiegel *et al.*, 2004; Johnson *et al.*, 2004; Turi and Kalen, 2000) with 24% occurring with lordosis (Johnson *et al.*, 2004), and 38 - 50% with spondylolisthesis (Spiegel *et al.*, 2004). In a population of patients with spastic diplegia aged 20 - 30+ years who did not undergo SDR, back pain was reported in 53 - 64% of the cohort (Harada *et al.*, 1993), and 55% of their patients with spondylolysis complained about back pain, while 75% with lordosis curves  $>70^\circ$  and 39% with curves  $<50^\circ$ . Our adults with spastic diplegia who did receive SDR had a higher frequency of back pain, but this was similar to adults who have not had SDR.

### **Methodological Limitations**

There are several potential limitations to our study. First, the sample size was relatively small, especially in the subgroup analysis; therefore, some statistically significant associations may not have been observed and these results should be regarded as preliminary. However, these are the only data on long-term spinal deformities after SDR. Second, there is limited data on long-term spinal deformity of patients with CP who did not receive SDR. We could not adequately compare our results with the natural history of the condition, and so cannot distinguish an independent effect of SDR. Third, the outcomes of the short-term follow-up were based on historical radiographic reports of analogue X-rays, while the long-term outcomes were evaluated from digital X-rays. Although there was a difference in type

of X-rays, we still believe that these results are reliable and the evaluations of both studies were completed by the same experienced neurosurgeon (JCP) and orthopaedic surgeon (EBH). Fourth, even though the follow-up period in the study is long, our patients were still relatively young (age range 21 - 44 years), and follow-up to later ages is still necessary.

### **CONCLUSIONS**

Seventeen to twenty-six years after SDR spinal deformities, except for spondylolisthesis, did appear to be more common. However, this increase was not marked. In fact, the only statistically significant increase was for relatively mild scoliosis. The majority had curves of less than 30 degrees and only 2 patients had curves of 35 degrees. No patient to date has required any spinal operations after their SDR. It is known that spinal deformities also increase as part of the natural history of CP, so there is a need for further studies to ascertain if the rate of development of spinal deformities after SDR is any different from that of the natural history of the condition.

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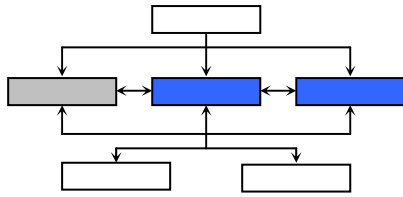
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# Chapter 6



Level of activity and participation in adults with spastic diplegia 17 to 26 years after selective dorsal rhizotomy

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## ABSTRACT

**Purpose:** To evaluate the activity and participation levels for adults with spastic diplegia 17 - 26 years after selective dorsal rhizotomy; to investigate relationships between subjects' characteristics and functioning; and their perceptions of the postoperative outcomes. **Method:** Thirty-one subjects with spastic diplegia, aged 21 - 44 years, who received rhizotomy between 1981 and 1991, participated in an observational follow-up study. A semi-structured interview was used to gather patients' characteristics and long-term experiences after the operation. The Functional Mobility Scale (FMS) and LIFE-H questionnaire were completed. **Results:** Based on FMS 84% were reported as independent for a distance of 5m, and 61% for 50 and 500m. Eighty percent were independent in accomplishing all life habits, with most problems found for Mobility and Recreation. This was in line with the subjects' perception, with strong correlations between LIFE-H accomplishment and satisfaction levels. Gross Motor Function Classification System levels were significantly correlated with the level of functioning, though age and socio-economic status showed no relationship. **Conclusions:** More than 15 years after rhizotomy, adults with spastic diplegia showed high levels of functioning, and similar levels of satisfaction with life habits. The majority had positive feelings about this neurosurgical procedure, although there is a need for better follow-up after school.

**Keywords:** Cerebral Palsy • Dorsal rhizotomy • Adults • ICF • Function • Participation • FMS • LIFE-H

## INTRODUCTION

Cerebral palsy (CP) is the most common cause of physical disability in childhood, with about 80% of children classified with the pyramidal (spastic) syndrome (Stanley *et al.*, 2000). CP cannot be cured, but biomedical, (re)habilitative and surgical interventions can prevent secondary abnormalities and improve functional abilities and quality of life (Rosenbaum and Stewart, 2004). One of the treatment options to ameliorate the patients from the disabling spasticity is the neurosurgical procedure known as selective dorsal rhizotomy (SDR).

SDR entails cutting a fraction of the nerve rootlets in the lower spine, thus interrupting the abnormal reflex circuit which gives rise to uncontrolled spasticity. This procedure was first described almost 100 hundred years ago by Foerster (1913), and was then further refined in the 1960's (Gros *et al.*, 1967) and 1970's (Fasano *et al.*, 1976). Peacock (Peacock and Eastman, 1981; Peacock and Arens, 1982) shifted the site of rhizotomy from the conus medullaris region to the cauda equina and reintroduced the technique in the 1980's. After the relocation of Peacock from Cape Town (South Africa) to Los Angeles (USA), SDR gained acceptance and is now used around the world (Hesselgard *et al.*, 2007).

The International Classification of Function, Disability and Health (ICF) model (World Health Organization, 2001) provides a biopsychosocial framework to identify and quantify the effects of interventions particularly in clinical trials (Brockow *et al.*, 2004; Stucki *et al.*, 2002; Stucki, 2005) and has been shown to be useful in the field of CP (Majnemer and Mazer, 2004; Rosenbaum and Stewart, 2004; Liptak, 2008). Based on the different domains of this model, Steinbok (2007) reported in his review article strong evidence of improvement in impairments and gross motor function and good evidence for benefits in performance of daily activities during the first few years after SDR.

In the literature most attention has been paid to the first decade of life in children with CP, despite the fact that CP is not life threatening and therefore children survive to become adults with persistent impairments of bodily structure and function as well as limitations in activity and participation levels (Klingbeil *et al.*, 2004; Liptak, 2008).

Long-term outcome studies are essential to guide future decisions in health care, education and social services (Majnemer and Mazer, 2004).

Our research team has reported on long-term outcomes in the domains of *Body Function & Structure* and the specific *Activity* of mobility in a test setting (capacity-level) (Langerak *et al.*, 2008; Langerak *et al.*, 2009a; Langerak *et al.*, 2009b). We identified a need to investigate the broader capacity of this population. Therefore, the question remains: What are the long-term outcomes of SDR in adults with CP for the domains of *Activity* (performance-level) and *Participation*?

Tools which comprehensively measure levels of performance in the domains of *Activity* and *Participation* were reviewed by Resnik and Plow (2009). They investigated 40 measures, and selected 5 questionnaires which linked questions to the 9 domains of the ICF-model: *Activity* and *Participation*. The Life-Habit questionnaire (LIFE-H) was one of the selected questionnaires documenting the manner in which people carry out daily activities and social roles (accomplishment level), and evaluates their perception regarding the level of participation achieved (satisfaction level), with the interaction of personal and environmental factors (Fougeyrollas *et al.*, 1998).

The quality of *Activity* performance in people with CP is often dependent on their functional mobility. Functional mobility can be defined as the manner in which people are able to move around in the environment in order to achieve daily activities and interact with society. Based on that concept, Graham *et al.* (2004) developed the Functional Mobility Scale (FMS). This scale differentiates itself from other activity limitation measures by the incorporation of the environment and the possibility of using different assistive devices (Harvey *et al.*, 2008).

A combination of the LIFE-H questionnaire and the FMS gives an in-depth insight into people's overall functioning. The aim of this study was to evaluate the activity limitations and restrictions in participation for adults with spastic diplegia, who had undergone SDR as children more than 15 years prior to this study. Secondary aims were developed to investigate possible relationships between sociodemographic, SDR and CP-related factors and these outcomes, and the relation between subjects'

functioning and their level of satisfaction. Therefore we studied: (i) subjects' functional mobility assessed by the FMS; (ii) quality of social participation based on LIFE-H 3.1. (accomplishment and satisfaction levels); (iii) associations between subjects' age at SDR, current age, socioeconomic status (SES) and Gross Motor Functional Classification System (GMFCS) levels with FMS and LIFE-H outcomes; (iv) association between subjects' LIFE-H accomplishment and satisfaction levels; and (v) the person's perceptions of the SDR experience from their current perspective.

## METHODS

### *Subjects*

Subjects were selected from a cohort of people with CP who underwent SDR at Red Cross Children's Hospital in Cape Town between 1981 and 1991. They were included if they were diagnosed with spastic diplegia, with or without mild unilateral upper extremity involvement, and excluded if they were diagnosed with dystonia, athetosis, ataxia or hypotonia. Historical clinical reports were used to confirm that the aim of the SDR procedure was functional improvement, to improve the gait pattern, or to develop locomotion if the child was not yet ambulant. In addition, the subjects had to live in Cape Town or within a radius of 100 kilometres at the time of data collection.

From the 106 subjects who underwent SDR in the specific time frame, 47 fulfilled the inclusion criteria from the clinical record audit. We had very limited contact details for these subjects, except for the 7 who had also participated in our completed 20 year follow-up studies (Langerak *et al.*, 2008; Langerak *et al.*, 2009a). A central CP register does not exist in South Africa, and so an intensive search based on peoples' names and date of birth was necessary. With assistance of family and teachers, use of old medical records and internet contact websites, we were able to locate 37 possible subjects (79%). Telephone contact was established, and we confirmed that 1 person had passed away (accident), and 5 people could not participate as they were unable to attend data collection sessions. Each of the 31 subjects who were

eligible and participated in the study completed an informed consent form. This study was approved by the Human Ethics Committee at the University of Cape Town.

### ***Outcome Measurements***

#### *Subject Characteristics*

A neurosurgeon (JCP) and physiotherapist (NGL) assessed the subjects and rated their Gross Motor Function Classification Scale (GMFCS) level (Palisano *et al.*, 2008; Jahnsen *et al.*, 2006) and if they were classified as spastic diplegia with or without mild unilateral upper limb involvement. In addition, the subjects were questioned (based on a semi-structured interview) with regards to socio-demographic and SDR-related topics. The socio-demographic data included: age, gender, current health status, health care visits, marital status (children), living situation, socio-economic status (SES), highest obtained degree in education, employment/student and main income. SES was estimated by housing density, which was calculated as the number of people living in the house divided by the number of rooms (excluding kitchen and bathroom) in the house (Micklesfield *et al.*, 2007). SDR-related characteristics included information about subjects' age at SDR, follow-up time and their experiences with this neurosurgical procedure. With regards to these SDR experiences the following questions were asked: (i) Do you think the SDR operation was worthwhile?; (ii) Looking back, what do you feel is the most important thing you would have done differently (something we clinicians can learn from)?; and (iii) If you had to make the decision to undergo the SDR operation now by yourself (after experiencing the impact of this on your life), would you do it?

#### *Functional Mobility Scale*

Together with the subject, one of the two physiotherapists (NGL or SH) completed the FMS. The FMS is an outcome measure to classify walking ability, where the possible use of an assistive device is taken into account. The scale is based on a six level ordinal grading system, ranging from 1 (use of wheelchair) to 6 (independent on all surfaces). Subjects are rated for three different distances; 5, 50 and 500m (Graham *et al.*, 2004). The FMS has been used in different studies (Svehlik *et al.*, 2008; Ma *et al.*, 2006) and proven to be valid and reliable in children with CP (Graham *et al.*, 2004; Harvey *et al.*, 2009). The validity and reliability of FMS has not

been tested in adults. However, its use seems to be feasible for all age groups and is currently used for studies with adults [personal communication, Adrienne Harvey].

### *LIFE-H questionnaire*

One of the two physiotherapists (NGL or SH) completed the LIFE-H 3.1 with each subject. Both strictly followed the published guidelines and filled in the answers themselves, while consulting the subject thoroughly. The LIFE-H questionnaire is based on a theoretical reference model called 'The Disability Creation Process' (DCP). In this model a disease (e.g. CP) is related to personal and environmental factors, which determine peoples' accomplishment and satisfaction level for different life-habits.

The LIFE-H questionnaire's measurement of accomplishment levels has been tested for validity and reliability in people of all ages (from children to older adults) and different diagnostic groups (CP (Donkervoort *et al.*, 2007; Donkervoort *et al.*, 2009; Lepage *et al.*, 1998b; Lepage *et al.*, 1998a), stroke (Desrosiers *et al.*, 2005; Rochette *et al.*, 2001), myotonic dystrophy (Gagnon *et al.*, 2006; Gagnon *et al.*, 2007), spinal cord injury (Noreau and Fougeryrollas, 2000) and older adults with disabilities (Levasseur *et al.*, 2004; Poulin and Desrosiers, 2009)). The LIFE-H 3.1 showed acceptable content and convergent validity with similar constructs of other questionnaires (Noreau *et al.*, 2007; Desrosiers *et al.*, 2003; Desrosiers *et al.*, 2004), and demonstrated moderate to excellent test-retest and intra- and inter-rater reliability (Noreau *et al.*, 2007; Noreau *et al.*, 2004; Gagnon *et al.*, 2006; Fougeryrollas *et al.*, 1998). The questionnaire has been used in different study cohorts of children (Lepage *et al.*, 1998b; Lepage *et al.*, 1998a; Noreau *et al.*, 2007) and adults (Donkervoort *et al.*, 2007; Donkervoort *et al.*, 2009) with CP. With regard to the satisfaction scale of the LIFE-H 3.1, Poulin and Desrosiers (Poulin and Desrosiers, 2009) reported high test-retest intra-class correlation coefficients. In addition, they emphasized that both the accomplishment level *and* the satisfaction level should be used for a correct interpretation of an individual's actual level of participation.

The LIFE-H is based on 77 life habits, which are related to Daily Activities (DA) and Social Roles (SR) items. DA comprises 6 categories (Nutrition, Fitness, Personal Care, Communication and Housing), while SR is subdivided into another 6 categories

(Responsibilities, Interpersonal Relationships, Community Life, Education, Employment and Recreation). Each of these 12 categories incorporates 4 to 8 different life habits. For each of the categories of the DA and SR items a weighted Accomplishment (Acc) and Satisfaction (Sat) score is calculated. The weighted Acc-score is a result of the subject's level of difficulty and need for assistance to accomplish the life habit (Table 6.1a), while the Sat-score is based on the subject's level of satisfaction to achieve the life habit in the specific manner as described for the Acc-score (Table 6.1b).

**Table 6.1a** Calculation weighted Accomplishment score.

<i>Acc-score</i>	<i>Difficulty level</i>	<i>Assistance Type</i>
9	No Difficulty	No assistance
8	No Difficulty	Assistive device or adaptation
7	With Difficulty	No assistance
6	With Difficulty	Assistive device or adaptation
5	No Difficulty	Human assistance
4	No Difficulty	Assistive device or adaptation <i>and</i> human assistance
3	With Difficulty	Human assistance
2	With Difficulty	Assistive device or adaptation <i>and</i> human assistance
1	Accomplished by a proxy	
0	Not accomplished	
N/A	Not applicable	

Formula weighted Acc-score:	$(\sum \text{Acc-scores} \times 10) : (\text{number of applicable life habits} \times 9)$
Range weighted Acc-score:	0 to 10
Acc-score: Accomplishment score.	

**Table 6.1b** Calculation weighted Satisfaction score.

<i>Sat-score</i>	<i>Satisfaction level</i>
10	Very satisfied
5	Satisfied
0	More or less satisfied
- 5	Unsatisfied
- 10	Very unsatisfied

Formula weighted Sat-score:	$(\sum \text{Sat-scores}) : (\text{number of applicable life-habits})$
Range weighted Sat-score:	-10 to 10
Sat-score: Satisfaction score.	

### **Statistical Analysis**

Descriptive statistical analyses were used to summarize subjects' characteristics. For the subjects' level of *Activity* and *Participation*, mean values and standard deviations of the FMS and LIFE-H questionnaire outcomes were calculated. To investigate the outcomes of the LIFE-H more specifically, we categorized the results of the accomplishment component, based on the mean weighted score in 3 levels: (i) score  $\geq 8.0$ : independent with no difficulties (with or without assistance); (ii) score 5 - 8: independent with difficulties (with or without assistance); and (iii) score  $\leq 5.0$ : dependent, since the life habit is not carried out by the subject or accomplished with human assistance. The outcomes of the satisfaction level were categorized into two levels: (i) score  $< 0.0$ : dissatisfied; and (ii) score  $\geq 0.0$ : satisfied.

Relationships between a number of the subjects' characteristics and the outcome variables were examined. Spearman rank correlation analyses were performed to determine the bivariate correlation between age at SDR, current age, and SES with 5FMS and 50/500FMS, as the data were non-parametric. Pearson correlation tests were used to examine the correlation of the first three subject characteristic variables with mean LIFE-H Accomplishment outcomes (Acc-DA and Acc-SR) and for the Satisfaction (Sat-DA and Sat-SR) correlations. The mean Acc-DA and Sat-DA was calculated by taking the average of the scores on the 6 constituent items on the subscale, and the mean Acc-SR and Sat-SR was calculated on the average of the scores on the 5 constituent items of the subscale. A Bonferroni corrected alpha-level of  $p \leq 0.008$  was used as a threshold for statistical significance. Analysis of Variance was applied to determine the relationship between GMFCS levels and the two LIFE-H Accomplishment (Acc-DA and Acc-SR) and Satisfaction outcomes (Sat-DA and Sat-SR). Pearson correlations were performed to demonstrate any correlations between Accomplishment and Satisfaction LIFE-H scores for the mean DA- and SR-scores and the mean of the total scores.

## RESULTS

### *Personal and SDR-related Characteristics*

Table 6.2 presents detailed characteristics of the 31 adults with spastic diplegia, who participated in the current study. They were 21 years or older and underwent SDR more than 17 years ago, and at the time of surgery 71% were between 2 and 7 years old. They were all ambulant, classified at GMFCS levels I-III and only 5 (16%) subjects had spastic diplegia with mild unilateral upper limb involvement. The majority of the study cohort had no other health issues. However, 1 subject was diagnosed with Chron's disease and another one with Graves' disease, 5 (16%) subjects had squint, 2 females (6%) had regular bladder infections and 1 subject took medication for high blood pressure. As a result of these problems (which were unrelated to SDR), 5 subjects (16%) visited a specialist and 3 (10%) a general practitioner, while 2 subjects (6%) had received treatment from a physiotherapist to release back pain during the previous month.

Fourteen subjects (45%) had a partner and 6 (19%) had (non-disabled) children. None of the study cohort stayed in an institution, but lived alone, with parents, partner or other housemates. Eight subjects (26%) had a low SES based on their housing density ratio  $>1.5$  (e.g. 5 people in a house with 3 rooms), and only 2 subjects (6%) had special adaptations in their houses. More than half of the study cohort had higher education such as an earned degree, while 1 subject was still studying at a university. He and another 21 subjects (71%) had a job, and their loan resulted in the main income, while 7 (23%) received a disability pension and 2 (6%) relied on the income of their partner or parents.

In response to the questions about the SDR operation, 23 subjects (74%) agreed that it had been worthwhile, while 7 (23%) answered that they 'didn't know', and 1 had no positive feelings about this. Ten subjects (32%) realized that they should have done more exercises or sport and 6 (19%) regretted that they did not or could not carry on with physiotherapy after school; 4 (13%) had no idea if they should have done something different, while 11 (35%) were happy with what they had done in the last years. Twenty-five of the study cohort (81%) responded that they would have

decided to undergo the operation now they could make independent decisions, 4 of them (13%) were not sure, while 2 (6%) disagreed.

**Table 6.2** Characteristics of subjects.

<i>Variable</i>	<i>Subjects</i>
Current age, years, median (range)	26.8 (21 - 44)
Age at SDR, years, median (range)	5.2 (2 - 27)
Follow-up time after SDR years, median (range)	21.3 (17 - 26)
SES <sup>φ</sup> , median (range)	1.0 (0.3 - 4.0)
GMFCS, <i>n</i> (%)	
Level I	15 (48)
Level II	11 (36)
Level III	5 (16)
Gender, <i>n</i> (%)	
Male	18 (58)
Female	13 (42)
Marital status, <i>n</i> (%)	
Single	17 (55)
Relationship	8 (26)
Married	6 (19)
Living situation, <i>n</i> (%)	
Alone	2 (6)
With (grand)parents	21 (68)
With partner (or others)	8 (26)
Highest obtained degree in education, <i>n</i> (%)	
Primary school (Grade 1 - 7)	1 (3)
Secondary school(Grade 8 - 12)	13 (42)
Higher education (university, technicon, courses)	17 (55)
Employment/student, <i>n</i> (%)	
Full-time job (>36 h/wk)	19 (61)
Part-time job	2 (7)
Unemployed	6 (19)
House keeping	1 (3)
Looking for job	2 (7)
Student	1 (3)
Main income, <i>n</i> (%)	
Paid job	22 (71)
Disability pension	7 (23)
Family income	2 (6)

<sup>φ</sup>SES: socioeconomic status is based on housing density (number of people: number of rooms in the house) (Micklesfield *et al.*, 2007).

### **Functional Mobility Scale**

For the distance of 5m the subjects were rated as follows: 9 subjects (29%) at level 6: independent on all surfaces; 17 (55%) at level 5: independent on level surfaces; 1 (3%) at level 4: use of stick(s); and 4 (13%) at level 3: use of crutche(s). Since the outcomes of FMS at 50 and 500m were similar in this study, we reported this variable as 50/500m FMS. For these distances, 7 subjects (22%) were rated at level 6, 12 (39%) at level 5, and 12 at level 3.

### **LIFE-H questionnaire**

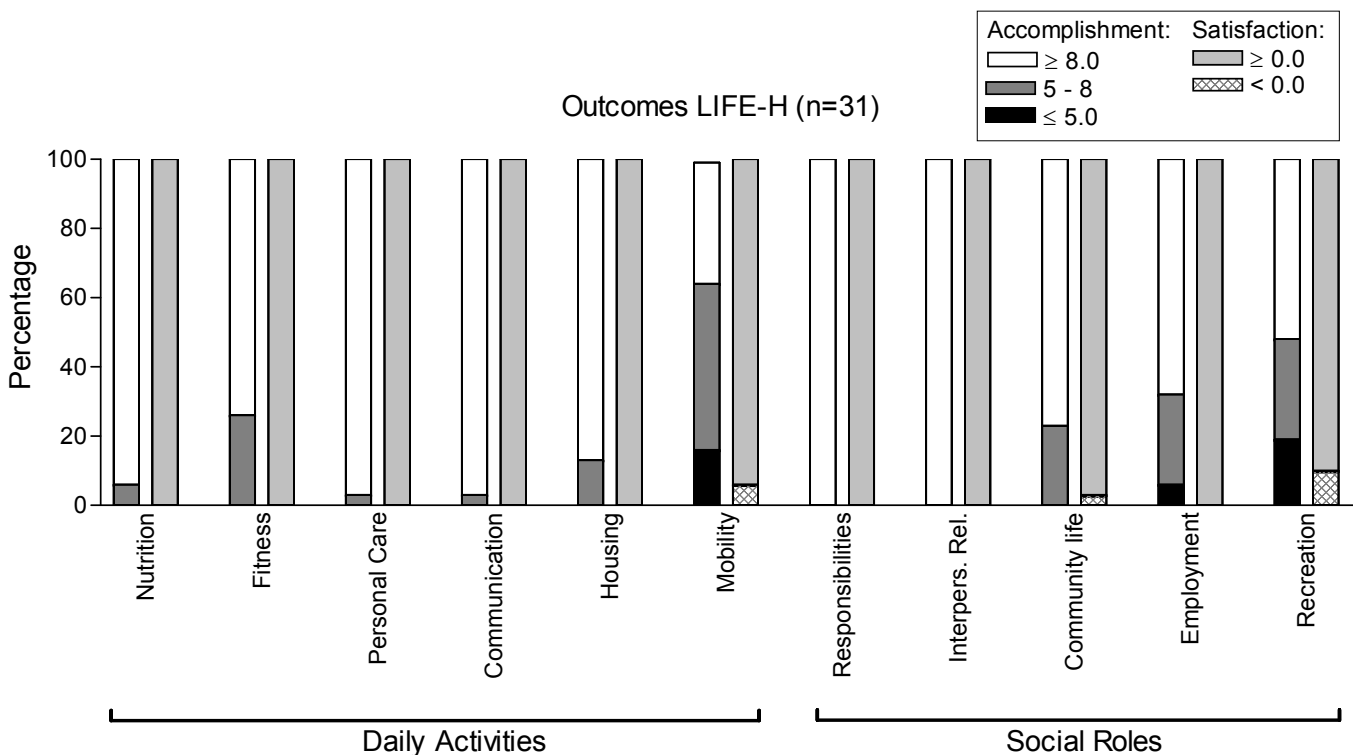
Table 6.3 shows the weighted mean scores with standard deviations and medians of the different categories of the LIFE-H questionnaire in regards to subjects' Accomplishment and Satisfaction levels. In addition, Figure 6.1 presents the percentages of subjects who were classified as independent with/without difficulties or dependent (accomplishment level) and satisfied or dissatisfied (satisfaction level) for the various daily activities and social role LIFE-H items.

**Table 6.3** Outcomes of the LIFE-H questionnaire.

<i>LIFE-H items</i>	<i>n</i>	<i>Accomplishment</i>			<i>Satisfaction</i>		
		<i>Mean</i>	<i>Median</i>	<i>SD</i>	<i>Mean</i>	<i>Median</i>	<i>SD</i>
Daily activities							
Nutrition	31	9.7	10.0	0.9	8.5	10.0	2.7
Fitness	31	9.0	9.7	1.3	7.4	7.5	2.6
Personal Care	31	9.7	10.0	0.5	8.5	10.0	2.1
Communication	31	9.8	10.0	0.5	9.0	10.0	2.0
Housing	31	9.3	9.6	1.0	8.0	10.0	2.7
Mobility	31	7.1	6.7	2.0	5.3	6.3	4.1
Total daily activities	31	9.1	9.2	0.7	7.8	8.8	2.3
Social roles							
Responsibilities	31	9.9	10.0	0.3	9.1	10.0	1.8
Interpersonal Relationships	31	9.8	10.0	0.5	8.7	10.0	2.5
Community life	31	8.9	9.4	1.5	6.8	8.6	3.9
Education <sup>‡</sup>	13	9.1	10.0	0.8	5.8	10.0	7.3
Employment	31	8.4	8.9	2.2	6.3	5.0	3.4
Recreation	30	7.4	8.6	2.8	5.6	6.4	4.7
Total social roles	31	8.9	9.2	1.1	7.3	7.9	2.6
Total	31	9.0	9.1	0.8	7.6	8.7	2.3

SD (standard deviation), <sup>‡</sup>Education (n = 13) was not included for calculation of Total social roles and Total scores.

With regard to daily activities, on average 81% of the subjects reported to be independent and had no difficulties to accomplish certain life habits. The subjects reported few problems with the items of Nutrition, Personal Care and Communication. Subjects faced the largest restrictions with the Mobility item. Sixteen percent of the subjects were dependent on human assistance or not able to accomplish the different mobility tasks at all, 48% were independent but had difficulties and 36% had no difficulties with this. In line with this, 6% of the study cohort reported they were dissatisfied with Mobility, while all subjects were (very) satisfied with how they accomplished the other daily activities. However, the variability of Mobility weighted scores was relatively large.



**Figure 6.1** Outcomes of LIFE-H questionnaire categorized. Accomplishment levels weighted score  $\geq 8.0$ : independent and no difficulties; score 5 - 8: independent with difficulties; and score  $\leq 5.0$ : dependent or not able to accomplish. Satisfaction levels with score  $\geq 0.0$ : satisfied; and score  $< 0.0$ : dissatisfied.

We excluded Education from the analyses of the 6 social roles, as this item was not applicable for the majority of the subjects. For the remaining 5 items, 79% of the subjects had on average no problems to accomplish these 5 SR items independently. The highest accomplishment levels were scored for Responsibility and Interpersonal Relationships, where none of them had any difficulties. This was in contrast to Employment and Recreation where relatively lower weighted scores were reported, with 6% and 19%, respectively, who did not accomplish this or only with human assistance, and also showed a greater variability compared to the other social role items. In regards to subjects' satisfaction levels, all subjects were satisfied with Responsibilities and Interpersonal Relationships, while 10% was not satisfied with their level of Recreation and 3% with Community Life. Despite the fact that some subjects had difficulties with Employment, none of them reported being dissatisfied in this domain.

### **Associations**

The estimated rank correlation coefficients as well as the  $p$ -values (two-tailed) regarding the relation between subjects' characteristic variables (age at SDR, current age and SES), and the FMS outcomes, and with the Life-H data (estimated correlation coefficients, not ranked) are shown in Tables 6.4 and 6.5 respectively. There were no significant relationship between any of the characteristics and any of the FMS or Life-H outcomes.

LIFE-H Accomplishment (Acc-DA and Acc-SR) and Satisfaction levels (Sat-DA and Sat-SR) were significantly related to subjects' GMFCS levels ( $p < 0.01$ ), except for Acc-DA and Acc-SR with GMFCS II and III (Figure 6.2). However it should be noted that GMFCS level III included only 5 subjects.

**Table 6.4** Spearman Rank Correlations between FMS and subjects' characteristic variables (n=31).

Variables	5 FMS		50/500 FMS	
	$r$	$p$	$r$	$p$
Age at SDR	0.23	0.22	0.27	0.14
Current age	0.26	0.17	0.14	0.46
SES	0.18	0.35	0.24	0.19

5FMS: FMS on 5 meter, 50/500FMS: FMS on 50 and 500m (50 and 500m were similar).  
P-values are two-tailed; significant at  $p \leq 0.008$ .

**Table 6.5** Pearson Correlations between LIFE-H and subjects' characteristic variables (n=31).

Variables	Acc-DA		Acc-SR		Sat-DA		Sat-SR	
	<i>r</i>	<i>p</i>	<i>r</i>	<i>p</i>	<i>r</i>	<i>p</i>	<i>r</i>	<i>p</i>
Age at SDR	0.02	0.91	-0.16	0.40	-0.16	0.38	0.09	0.59
Current age	0.16	0.38	-0.10	0.58	-0.06	0.74	0.24	0.19
SES	0.29	0.11	0.11	0.56	-0.29	0.64	0.08	0.67

Acc-DA: Accomplishment score for Daily Activities; Acc-SR: Accomplishment score for Social Roles; Sat-DA: Satisfaction score for Daily Activities; and Sat-SR: Satisfaction score for Social Roles. P-values are two-tailed; significant at  $p \leq 0.008$ .

A Pearson correlation analysis demonstrated a strong positive correlation between the total Accomplishment LIFE-H score and the total Satisfaction LIFE-H score,  $r=0.78$ ,  $p < 0.01$  (*two-tailed*), which is based on daily activities and social roles outcomes. Therefore, strong positive correlations were demonstrated between Acc-DA and Sat-DA and Acc-SR and Sat-SR ( $r=0.79$ ,  $p < 0.01$  and  $r=0.73$ ,  $p < 0.01$ , respectively).

## DISCUSSION

As CP is the result of a *non-progressive* injury to the developing brain (Rosenbaum *et al.*, 2007), it could be assumed that the short-term benefits from SDR as demonstrated by many studies (Steinbok, 2001), are maintained with aging. However, it has been shown that as people with CP who did not receive SDR become older, they can experience various secondary complications, with limitations on activity levels and negative influences on their quality of life (Klingbeil *et al.*, 2004; Liptak, 2008). In line with our previous long-term follow-up studies regarding adult impairments and functioning (capacity level) (Langerak *et al.*, 2008; Langerak *et al.*, 2009b; Langerak *et al.*, 2009a), this study also demonstrated the important benefits of SDR, but this time specifically in subjects' functional performance level and participation in the community more than 15 years after SDR.

### **Functional Mobility Scale**

Considering the outcome measures of the ICF-model component Activities, Bjornson (2008) suggested there is a difference between functional performance and

capability. Hulsebeeke et al. (2009) subdivided the level of functioning even further. They showed there is a difference for children with CP in their capacity (“can do in a standardized, controlled environment”), capability (“can do in daily environment”) and performance (“does do in daily environment”) levels. The current study focused on the subjects’ functional *performance* level.

The FMS was used as an outcome measure of subjects’ mobility levels in a daily indoors and outdoors setting. More than 15 years after SDR 84% of our study cohort was still able to walk for 5m independently (score 5 or 6 on FMS), and 61% of the subjects for 500m, while the remainder had to use sticks or crutches (score 3 or 4). This is in line with the outcomes of our previous 20 year follow-up study (Langerak *et al.*, 2007), where we reported that in general, 79% of the adults with spastic diplegia walked without assistive devices, 14% with crutches or cane and 7% became dependent on a wheelchair. These percentages of independent walking ability are higher than those reported for adults with spastic diplegia who did not receive SDR. Andersson and Mattsson (2001) showed that 49% walked without and 30% with walking devices, and 21% were not able to walk. Similarly, Ando and Ueda (2000) reported an incidence of 57% walking without and 43% with aids. However, it has to be taken into account that the average age where functional deterioration has been reported is about 37 years (Ando and Ueda, 2000; Opheim *et al.*, 2009), and that the age of our study cohort ranged from 21 - 44 with a median age of 27 years, while the mean age in the other two studies was mid 30’s and the oldest participants were in their 50’s.

Jahnsen et al. (2004) found that increased age was significantly correlated with a deterioration of locomotion skills. In addition, in a 7 year follow-up study of the same study cohort, they reported that 71% of the included adults with spastic diplegia presented deteriorated walking (Opheim *et al.*, 2009). We found no significant correlation between age and walking ability. No associations were found for FMS and SES either. Therefore we agree with Ando and Ueda (2000) that functional status may be determined by factors intrinsic to CP. However, we also believe that environmental factors play an important role in the development of patients’ functional level and should be included in any evaluation and clinical interpretation of patient status.

### ***LIFE-H questionnaire***

#### *Accomplishment with Daily Activities*

Based on the LIFE-H 3.1, we found that 81% of our study cohort had a mean LIFE-H Acc-DA score  $\geq 8.0$ , which means that 81% were independent and had no difficulties in accomplishing daily activities. Donkervoort et al. (2007), who published a study on people with CP aged between 16 - 20 years, reported similar scores for 79% of their study cohort, while they found 91% of their subjects were independent based on the Functional Independent Measure/Functional Assessment Measure (FIM/FAM). Andren & Grimby (2004) also used the FIM in a 5 year follow-up study of adults with CP and Spina Bifida, aged 24 - 43 years, and also found that the majority of their study cohort was independent, although there were some changes seen during the adult years. Van der Dussen et al. (2001) studied the functional level of adults with CP, aged 21 - 31 years, with the Barthel Index, and concluded that 75% of their subjects were independent for daily activities.

In other words, people with spastic diplegia (with and without minor involvement of unilateral upper limb) have a similar level of independency in daily activities compared to other adults with CP. This is also apparent when we compare our mean LIFE-H 3.1 Acc-DA scores with the outcomes of Donkervoort et al. (2007), which were 9.1 and 8.2 respectively. In addition, our Acc-DA score was relatively high compared to the outcomes of children with CP, aged between 5 - 17 years, as reported by Lepage et al. (1998b) They found median Acc-DA scores between 2.5 and 5.5 (with a very wide variability) based on the LIFE-H version 1.0.

More specifically, in our study cohort the greatest difficulties (highest level of dependency) for the Acc-DA was seen in the Mobility category. Sixty-four percent of our subjects had difficulties accomplishing life habits related to mobility (Acc-score:  $< 8.0$ ). However, 48% were still able to accomplish the life habits independently (Acc-score: 5 - 8), while 16% either did need human assistance or did not accomplish these tasks (Acc score:  $\leq 5.0$ ). Lepage et al. (1998b) found the most problems in the Community category, although their median Acc-DA score for Mobility was still much lower than ours, estimated as 4.2 compared to our 7.1. Donkervoort et al. (2007) also reported for their 103 adults with CP, who did not receive SDR, the lowest mean

Acc-DA scores for the Mobility category, with 27% of their study cohort having difficulties and a mean score of 7.9. This score was not especially different to our outcome, while the percentage of subjects with Mobility difficulties was much higher. This latter fact can be explained by a much greater variability (standard deviation) in our study cohort, which might be due to an older and wider age range. However, we found no significant correlations between age and the LIFE-H Acc-DA scores. In addition, no significant correlations were found with age at SDR.

### *Accomplishment with Social Roles*

In regards to the accomplishment of LIFE-H social roles we found, similar to Acc-DA, a mean LIFE-H Acc-SR score  $\geq 8.0$  in 79% of our study cohort. This was just higher than the 76% reported by Donkervoort et al. (2007) in adolescents and young adults with CP, who did not receive SDR. The mean Acc-SR scores of 8.9 and 8.2, respectively, as reported in current study and that of Donkervoort et al., were also similar. However, these scores were much higher than those reported in children with CP, with median scores ranging from 0.5 - 6 (Lepage *et al.*, 1998b).

We found in our study cohort that most subjects had no difficulties with the Acc-SR categories Responsibilities and Interpersonal Relationships, but more problems were reported in Employment and Recreation. If we compare these outcomes with the study of Donkervoort et al. (2007), the number of subjects who had difficulties with the Employment category are similar, but their study cohort had apparently fewer problems accomplishing the Recreation tasks. Forty-eight percent of our South African group reported having difficulties with this SR category, while a score  $< 8.0$  was found in 27% of the Dutch group. It might be possible that the explanation lies in the differences between the countries; although South Africa is a developed country, the facilities for people with disabilities are not as well organized as in the Netherlands.

However, we found no statistically significant relationship between SES and mean Acc-SR scores. In addition, there were also no correlations found with current age, age at SDR, but there was a highly significant correlation with GMFCS which will be discussed below.

### *Satisfaction with Life Habits*

In addition to the Accomplishment component, the LIFE-H questionnaire also consists of a Satisfaction component. Unfortunately this part has not been reported often in the literature, despite the perception of the patient, in addition to the accomplishment of life habits, being acknowledged as such an important component of outcome measurement (Noreau *et al.*, 2004; Poulin and Desrosiers, 2009). In addition, it has been shown that, in older adults with physical disabilities, LIFE-H Satisfaction scores were more strongly correlated to peoples' quality of life than their Accomplishment scores (Levasseur *et al.*, 2004).

With regard to LIFE-H 3.1 Sat-DA categories, the subjects of our study cohort reported being (very) satisfied, except for 6% for the Mobility category. Based on the questions about Social roles, subjects were in general satisfied with how they achieved the described life habits. However, as described for Acc-DA, most problems were found in the Recreation category with 10% reporting a negative mean Sat-SR score, which suggests that they were unsatisfied. In line with the LIFE-H accomplishment items, no significant correlations between patient characteristic variables and LIFE-H Sat-DA and Sat-SR were found, except for subjects' GMFCS levels.

### **Associations**

In this current study strong statistically significant correlations for the total LIFE-H Accomplishment and Satisfaction levels ( $r=0.80$ ) were found, and also for the subscales of total DA ( $r=0.82$ ) and SR ( $r=0.66$ ) scores. This is in contrast to the recently published study of Poulin and Desrosiers (2009), where correlations of 0.43 for the averages of all categories were reported, and 0.36 for DA and 0.24 for SR. They used the LIFE-H questionnaire in a population of older adults with functional disabilities, aged  $\geq 65$  years, which might be the reason for this apparent difference.

As also presented in previous studies focusing on children, adolescents and young children with CP (Donkervoort *et al.*, 2007; Beckung *et al.*, 2007), gross motor functioning levels as defined by the GMFCS differentiate outcomes of functioning. Bagley *et al.* (2007) studied the discrimination levels of different outcome tools associated with the *Activity* and *Participation* components of the ICF-model. They

concluded that GMFM-66, velocity and WeeFIM were the best discriminatory tools for GMFCS levels I and II, and GMFM Dimension E, Pediatric Functional Independence Measure (WeeFIM) Self-Care and Mobility, cadence, and Gillette Functional Assessment Questionnaire Question 1 for GMFCS levels II and III, while Quality of Life and cognition measures were the least discriminative tools. Unfortunately the LIFE-H was not evaluated in that paper, but should be of serious interest for future investigations as it might discriminate between each of the three GMFCS levels and covers the *Activity* as well as the *Participation* component of the ICF-model.

### ***SDR Experience***

In line with the high level of satisfaction as reported for the LIFE-H questionnaire life habits by our subjects more than 15 years after SDR, the majority of this study cohort reported that they found the SDR operation worthwhile and would have elected to receive the operation if they had to decide themselves. However, more than half of the cohort reported that there was a lack of physical activity and physiotherapy after they left school. The lack of care with transition into adulthood has also been described in other studies, and different research groups have emphasized the demand for better long-term follow-up with guidance, training programmes and health care after leaving school (Andersson and Mattsson, 2001; Jahnsen *et al.*, 2004).

### ***Limiting Factors***

One of the aspects that has to be noted is that our sample size of 31 subjects was relatively small, meaning that our outcomes should be seen as an important indication of long-term outcomes, but not as definitive. In addition, this study was conducted in South Africa, and although it is a developed country, the health environment is not as well developed as in Western countries. We also do not know if the cultural differences have an influence on peoples' level of coping, which in turn might be related to patients' functioning and satisfaction in life.

## **CONCLUSIONS**

With this first published long-term follow-up study we can conclude that adults with spastic diplegia (with or without mild involvement of unilateral upper limb) and who

have undergone SDR as children, have a high level of functioning within the *Activity* and *Participation* components of the ICF-model. The levels of functioning as measured with the FMS and LIFE-H questionnaire are related to GMFCS levels I, II and III. In addition, subjects' accomplishment levels are strongly correlated with their levels of satisfaction, which is in line with their positive feelings about the SDR operation. However, it has to be noted that there is a great demand for better and more care as people with CP make the transition from adolescence to adulthood.

## ACKNOWLEDGEMENTS

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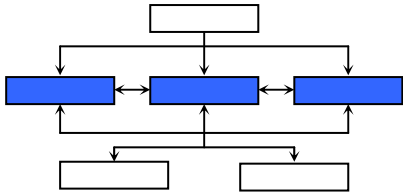
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# Chapter 7

Summary and Conclusions

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## **RATIONALE OF THE THESIS**

Cerebral Palsy (CP) is known as a leading cause of disability in childhood, with a prevalence of about 2.0 - 2.5 per 1000 live births (Odding *et al.*, 2006; Stanley *et al.*, 2000a). Although CP results from a *non-progressive* disturbance to the developing brain (Rosenbaum *et al.*, 2007), it is a complex condition with various secondary complications and accelerated impairments occurring with age (Liptak, 2008; Roebroek *et al.*, 2009; Klingbeil *et al.*, 2004; Rapp and Torres, 2000).

About 80% of the patients with CP are diagnosed with the spastic type (Stanley *et al.*, 2000b), and it is important to treat the spasticity with insight. There are various treatment options available, ranging from physical therapy, device-assisted modalities, oral and parenteral medication through to surgery (Goldstein, 2004; Rosenbaum, 2003; Sussman and Aiona, 2004; Koman *et al.*, 2004; Krigger, 2006; Steinbok, 2006; Tilton, 2009). Selective dorsal rhizotomy (SDR) is a neurosurgical procedure, which is an irreversible intervention for focal spasticity mainly in the lower extremities. The operation has a particular appeal to interrupt the abnormal reflex circuit that maintains spasticity (Foerster, 1913).

SDR had limited acceptance until it was refined and reintroduced by Professor Warwick Peacock at Red Cross Children's Hospital, Cape Town, in the early 1980's (Peacock and Eastman, 1981; Peacock and Arens, 1982). During the last few decades different variations of this technique have been proposed (Steinbok *et al.*, 2009; Park *et al.*, 1993; Cochrane and Steinbok, 1992; Kim *et al.*, 2002), but the 'Peacock method' is currently still the most widely used worldwide (Hesselgard *et al.*, 2007). A large number of studies have demonstrated the benefits of this neurosurgical procedure (Steinbok, 2001; Farmer and Sabbagh, 2007), but these conclusions were based on assessments performed within the first few years after surgery.

There is thus a need for long-term outcomes (van Schie *et al.*, 2005; Farmer and Sabbagh, 2007) which will help decision making for clinicians, parents and patients. Since the 'Peacock method' for SDR was reintroduced here in Cape Town, we are the first research group who are in a position to provide the long-term outcomes.

The aim of this doctoral thesis was therefore to investigate the long-term sequelae of SDR in patients with spastic diplegia seen in the context of the broad spectrum of the ICF-model: *Body Function & Structure, Activities and Participation*.

**Table 7.1** Constructs for ICF-model components.

<i>Body Function</i>	<i>Body Structure</i>	<i>Activity and Participation</i>
b1. Mental functions	s1.* Structure of the nervous system	d1.* Learning and applying knowledge
b2.* Sensory functions and pain	s2. The eye, ear and related structures	d2.* General tasks and demands
b3. Voice and speech functions	s3. Structures involved in voice and speech	d3.* Communication
b4. Functions of the cardiovascular, hematological, immunological and respiratory systems	s4. Structure of the cardiovascular, immunological and respiratory systems	d4.* Mobility
b5. Functions of the digestive, metabolic and endocrine systems	s5. Structures related to the digestive, metabolism and endocrine systems	d5.* Self care
b6. Genitourinary and reproductive functions	s6. Structure related to genitourinary and reproductive system	d6.* Domestic life
b7.* Neuromusculoskeletal and movement related functions	s7.* Structure related to movement	d7.* Interpersonal interactions and relationships
b8. Functions of the skin and related structures	s8. Skin and related structures	d8.* Major life areas d9.* Community, social and civic life

\*These constructs have been addressed in this thesis.

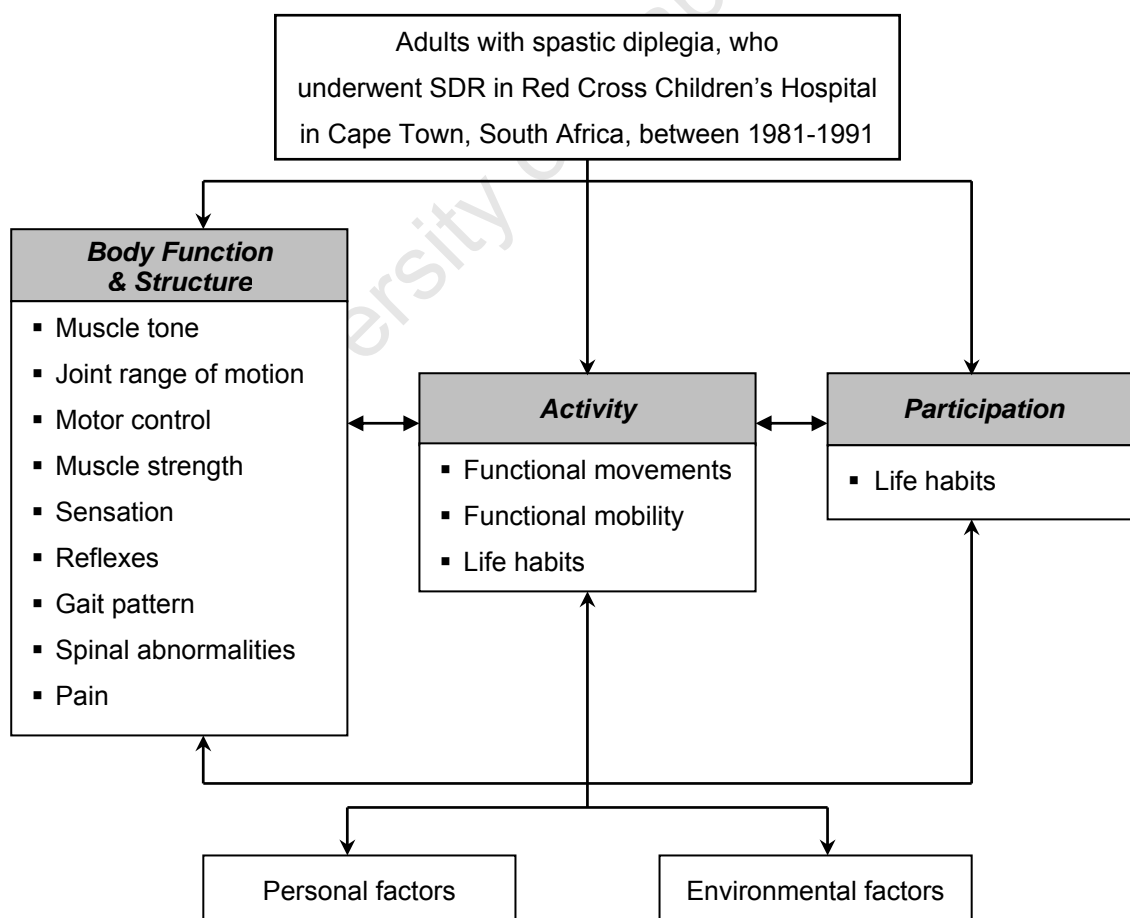
### ***ICF-model***

To acquire a clear overview of the long-term sequelae of SDR, we have applied the International Classification of Functioning, Disability and Health (ICF) model developed by the World Health Organization (WHO, 2001). As described in Chapter 1 (Figure 1.2), the ICF-model includes two components: (i) *Body Function & Structure*; and (ii) *Activities and Participation*. These two components are divided in different constructs as shown in Table 7.1. For our patients with spastic diplegia who

underwent SDR, we focused on the four most relevant constructs from the *Body Function & Structure* component: (b7) neuromusculoskeletal and movement related functions; (b2) pain and sensation; (s1) structure of nervous system; and (s7) structures related to movement. For the *Activity* and *Participation* component, we concluded that each construct would be worth studying (d1-d9), with some extra emphasis on mobility (d4).

### Research Projects

The different aspects of the ICF constructs chosen for our research projects are presented in Figure 7.1, and an overview of the projects and chapters wherein these were discussed is given in Tables 7.2 and 7.3. The identification of the aspects was in the first place based on our earlier experiences with SDR, which were described in the first article of this thesis (Chapter 2) and will be referred to as project 0. Chapter 2 also includes some preliminary data of our first long-term follow-up project (project I).



**Figure 7.1** The ICF-model (WHO, 2001) with identification of specific aspects addressed in thesis.

Project I was conducted in 2005. We performed two 20 year follow-up studies with 14 patients with spastic diplegia, focusing on 2-dimensional gait analysis (Chapter 3) and also on neuromuscular and functional outcomes (Chapter 4). These studies provided scientific evidence for the long-term positive benefits of SDR. However, this data also highlighted some concerns about potential spinal instability and we acknowledged methodological limitations such as sample size, validated outcome measures and the lack of a control group. Except for including a ‘true’ CP control group (age matched patients with spastic diplegia who did not receive SDR), the other problems were taken into account for our second project.

**Table 7.2** Aspects of thesis related to ICF-model component *Body Function & Structure*.

<i>Topics</i>	<i>Project</i>	<i>Chapter</i>	<i>Outcome measure</i>
Muscle tone	0	2	Description of Cape Town experiences
	I	4	Muscle tone scale (Berman, 1989)
	II	5	Ashworth scale (Ashworth, 1964)
Joint Range of motion	0	2	Description of Cape Town experiences
	I	3	Gait analyses
	I	4	Joint stiffness scale (Berman, 1989)
Motor control	0	2	Description of Cape Town experiences
	I	4	Voluntary movement scale (Berman, 1989)
Muscle strength	0	2	Description of Cape Town experiences
	I	4	Voluntary movement scale (Berman, 1989)
	II	5	MRC-scale (WHO, 1995)
Gait pattern	0	2	Description of Cape Town experiences
	I	3	Gait analyses
Sensation	II	5	Pin-prick, touch and pressure assessments
Reflexes	II	5	Deep tendon and Babinski
Spinal abnormalities	0	2	Description of Cape Town experiences
	II	5	X-ray and MRI-scans
Pain	II	5	ODI and general questionnaire

ODI: Oswestry Disability Index (Fairbank *et al.*, 1980).

**Table 7.3** Aspects of thesis related to ICF-model component *Activity* and *Participation* component.

<i>Topics</i>	<i>Project</i>	<i>Chapter</i>	<i>Outcome measure</i>
Functional movements	0	2	Description of Cape Town experiences
	I	4	Functional movement scale (Berman, 1989)
Functional mobility	I	4	GMFCS (Palisano <i>et al.</i> , 1997)
	II	6	FMS (Graham <i>et al.</i> , 2004)
Life habits	I	4	Self-developed questionnaire
	II	6	LIFE-H 3.1 (Fougeyrollas <i>et al.</i> , 1998)

GMFCS: Gross Motor Function Classification System; FMS: Functional Mobility Scale; LIFE-H 3.1: LIFE-Habit 3.1 questionnaire.

Data collection for project II took place in 2008. We tracked down 32 adults with spastic diplegia who received SDR between 1981 and 1991 and were willing to participate in this second research project. These patients received X-rays and MRI-scans of their spine, were clinically assessed by a neurosurgeon and physiotherapist, and completed a general questionnaire, and also a questionnaire related to back pain and life-habits (Chapters 5 and 6).

### ***Patient Cohorts***

The patients who participated in both projects were diagnosed with spastic diplegia, while 5 patients who participated in project II also had some minor involvement of unilateral upper limb. The distribution of female:male was about 4:6, and they had a mean age of approximately 27 years (range 21 - 44 years). Patients were classified at GMFCS levels I-III except for 1 patient at GMFCS level IV (project I). Patients were relatively healthy, with no sexual, bowel or bladder function problems related to SDR. However, one patient who participated in project II was diagnosed with Crohn's disease and another patient with Graves' disease. None of the participants had ever received botox, intrathecal baclofen or any spinal interventions, while about half of the cohort underwent orthopaedic surgery before and/or after SDR. Patients had a different socio-economic status, but all of them had attended primary school, and more than three-quarters of the study cohort were employed or students at the time of data collection.

## **CONCLUSIONS OF STUDIES**

### **Body Function & Structure**

During the 1980's and 1990's the Cape Town research team conducted different follow-up studies and provided evidence for the benefits gained as a result of the SDR operation (Chapter 2). The one year follow-up studies by Berman (1989) showed positive outcomes for the parameters muscle tone, joint stiffness (range of motion) and voluntary movements (combination assessment of muscle strength and motor control). Peter *et al.* (1990; 1993) focused on spinal deformities and reported that only the incidence of spondylolysis was significantly higher compared to patients with CP who did not receive SDR. Vaughan *et al.* (1988; 1991) focused on gait analyses, and showed the biggest changes during the first three years after SDR. However, 10 years postoperatively they still reported significant improvements in hip and knee range of motion, which also resulted in improved walking speed due to increased step length (Subramanian *et al.*, 1998).

In the long-term some deterioration was shown based on 2-dimensional gait kinematic and temporal-distance parameters. However, 20 years after SDR we still showed gains in patients' gait compared to preoperative patterns, (Chapter 3). Gait patterns are a result of patients' muscle tone, joint range of motion and motor control. More than 15 years after SDR patients still had reduced muscle tone, approaching normal values (Chapters 4 and 6). In addition, active (based on gait analyses) and passive (based on joint stiffness assessments) joint range of motion did improve during the years after SDR (Chapters 3 and 4).

It is suggested that muscle strength rapidly deteriorates during the first years after SDR, which result in a collapsed gait pattern with increased hip and knee midrange values (Chapter 3). However, after intense physiotherapy patients do recover, and in the long-term more than half of the study cohort had normal muscle strength with otherwise most impairments seen in the hip extensors and knee flexors (Chapter 5). A combination of muscle strength and motor control was tested by voluntary movement assessments. Our long-term follow-up study showed significant improvement for this parameter 20 years after SDR compared to before surgery (Chapter 4).

Other clinical assessments showed that the majority of patients had no impaired pin-prick sensations, while none of them had abnormalities in proprioception, touch or deep pressure. In addition, most patients had reduced deep tendon reflexes and positive Babinski reflexes (Chapter 5).

With regards to spinal deformities, the incidence of spondylolisthesis did not progress in the long-term, while scoliosis, kyphosis, lordosis and spondylolysis were more common 17 - 26 years after SDR. However, it was only the incidence of scoliosis that was significantly increased, with only 7% of patients having a curve  $>30^\circ$ . In addition, no high incidences of degenerative disorders (e.g. spinal stenosis, black discs, and disc herniation) or other spinal abnormalities were found, while back pain on a daily basis was reported in 17% of the study cohort (Chapter 5).

### ***Activity and Participation***

Patients' level of *Activity* can be quantified in a test setting or in daily environment, which should be defined as the patients' capacity and performance level, respectively. Based on functional movement assessments (long sitting, side sitting, half kneeling, prone kneeling, kneel standing, rolling, standing, crawling and walking), improvements were seen in patients' capacity level 1 year (Chapter 2) and 20 years after SDR (Chapter 4).

These gains were also reflected in patients' performances in daily life. Although the Gross Motor Function Classification System (GMFCS) was developed as a classification system and not as an outcome measure, we showed a positive change in 64% of our study cohort who participated in project I, 29% stabilized and 7% deteriorated over time. This means that 20 years after SDR, 79% of the patients walked without assistive devices, 14% walked with a crutch or cane and 7% used a wheelchair (Chapter 4). Based on the Functional Mobility Scale (FMS) we established that 84% of the patients who participated in project II could walk without any assistive devices for 5 metres and 61% for 50 and 500 metres more than 15 years after SDR (Chapter 6). In relation to back pain and lower extremity pain, 77% of the study cohort faced no or minor disability, while 23% experienced 'moderate disability' (Chapter 5)

When we asked the patients who participated in project I, “Do you need help with activities of daily living?” all participants answered “no” to the question (Chapter 2 and 4). When we addressed this question in a more objective way, by using the LIFE-H questionnaire 17 - 26 years after SDR, 80% of our study cohort reported to be able to accomplish all different life habits independently and without any difficulties. These life habits were related to (i) Daily activities: Nutrition, Fitness, Personal Care, Communication, Housing, Mobility; and (ii) Social roles: Responsibilities, Interpersonal Relationships, Community life, Employment and Recreation. Most problems were found with the Mobility and Recreation items with 36% and 52% respectively of the study cohort who were independent and had no difficulties to accomplish these items (Chapter 6).

### ***Experiences of Patients***

In general, patients were satisfied with the way they were able to achieve their life habits 17 - 26 years after SDR. We found a strong correlation between the LIFE-H accomplishment and satisfaction levels for daily activities and social role life habits. Ninety-eight percent of the patients were either satisfied or very satisfied with their life habits. Again most problems were seen for Mobility and Recreation, with 6% and 10% respectively being dissatisfied or very dissatisfied with the way they are able to accomplish these activities.

The majority of patients who participated in both our projects had positive feelings about the SDR operation. The exceptions were 2 patients in project I and 1 patient in project II. The major problem appeared to be muscle weakness postoperatively despite intensive physiotherapy both pre-and postoperatively.

In addition, we asked the patients in project II, “Looking back, what do you feel is the most important thing you would have done differently (something we clinicians can learn from)?”. Thirty-two percent of the study cohort responded that they should have done more exercises or sport and 19% regretted that they did not carry on with physiotherapy after school. The last question we asked was “If you had to make the decision to undergo the SDR operation now by yourself, after experiencing the impact of this on your life, would you do it again?”. Eighty-one percent of the study

cohort responded that they would undergo the operation now they could make an independent decisions, 13% were not sure, while 6% disagreed.

## OVERALL CONCLUSIONS

More than 15 years after SDR we are able to provide an indication of positive outcomes for the SDR operation with regards to the ICF-model components *Body Function & Structure* and *Activity and Participation*. Despite the small sample sizes of our study cohorts, it is apparent that the patients with spastic diplegia gained sustained benefits with regards to muscle tone, motor control and muscle strength as demonstrated by improvements in their gait patterns. Disturbed pin-prick sensation, reduced reflexes, some spinal abnormalities and an increase of spinal deformities incidences were reported. However, no specific concerns regarding spinal abnormalities were reported. Only the number of patients with scoliosis did increase significantly, but this included mainly curves  $<30^\circ$ . Low back pain was indicated, but this did not lead to disability in daily activities for the majority of patients.

Level of *Activity and Participation* of the patients was relatively good, with most of them being independent and experiencing no problems with accomplishing various life habits. Mobility was seen as the life habit which resulted in most problems, despite the fact that after SDR positive changes in GMFCS levels were seen and the majority walked independently as indicated by FMS score 5 and 6. Mobility was also the life habit item that patients were less satisfied with, which confirms the strong relationship between patients' level of accomplishment and satisfaction in life habits. The majority of the patients had positive feelings about the SDR operation, but they reported they would have liked more exercises and physiotherapeutic guidance after leaving school.

We would like to emphasize that strict selection criteria are necessary, long-term follow-up studies and guidance are important, and further studies are still necessary. Such studies will enable researchers to be able to make more definitive conclusions, to explore mobility outcome measures over a life-span, to address relations with contextual factors, and to evaluate conformity with other countries.

## RECOMMENDATIONS

### ***Patient Selection***

As mentioned in the introduction to this thesis (Chapter 1), strict selection criteria are very important, and we would like to emphasize again that this is the key factor for success with SDR. Besides the specific clinical factors, the child and parents (or other caregivers) have to be motivated and eager for this invasive neurosurgical procedure *with* intense rehabilitation postoperatively. A good understanding of the specific motor problems, the operation and expectations by the child and parents (or other care-givers) are very important and should not be underestimated when patients consult specialist clinicians.

### ***Guidance and Follow-up***

Based on our long-term follow-up studies we would like to emphasize that follow-up of patients is still very important. Standard follow-up programmes after children leave school should be developed, which should include clinical check-ups including X-rays of the spine, and a possibility for questions by clinicians with specific knowledge about the transition from childhood into adolescence and adulthood. In addition, patients should be stimulated to participate in sports or a physiotherapy programme, to carry on with an active lifestyle as promoted at school.

## FUTURE STUDIES

### ***Study Cohorts***

The sample sizes of our study cohorts were relatively small; in project I 14 patients participated, and project II was comprised of 32 patients. Both study cohorts were tested with different outcome measures (Tables 7.2 and 7.3), and therefore it was not feasible to combine the outcomes of the different aspects of the ICF-model constructs. Additional long-term follow-up measures would be of great value to assure the outcome of our projects.

In our follow-up studies we should have ideally included a control group of patients with CP who did not undergo SDR in the 1980's. Unfortunately, this was not ethically

possible, and therefore no 'true' control group existed. There is in general a shortcoming of long-term outcome studies based on adults with CP. Therefore we would like to emphasize that there is a great demand for studies with adults with spastic diplegia who did not undergo SDR (and preferably no other intervention), and adults who received other interventions in childhood to release them from spasticity or secondary abnormalities (e.g. intrathecal baclofen, botox, orthopaedic surgery).

In addition, it would be interesting to find out what the long-term outcomes are of patients who underwent an adapted SDR technique instead of the 'Peacock method', e.g. laminotomies instead of laminectomies (Cochrane and Steinbok, 1992), laminectomies caudal to the conus medullaris (Park *et al.*, 1993; Park and Johnston, 2006), with or without electromstimulation during the procedure (Steinbok and Kestle, 1996).

### ***Mobility Outcome Measures***

As can be inferred from the definition of cerebral palsy, "CP describes a group of permanent disorders of the development of movement and posture" (Rosenbaum *et al.*, 2007), functional mobility is an important aspect in the development of a child with CP. In addition, we found that in the long-term after SDR, mobility was one of the life habits where patients had most difficulties and were least satisfied. Therefore mobility is an important aspect that needs attention through patients' life-span. Seen in a broad spectrum of the ICF-model, each of the constructs for *Body Function & Structure* component (b7 and s7) and *Activity* and *Participation* (d4) should be taken into account. In line with this, we should in the first place suggest for future studies the use of 3-dimensional gait analyses instead of 2-dimensional analyses (we had no choice in our long-term follow-up study, since this was not available in 1985). Secondly, the FMS and LIFE-H outcome measures should be specifically validated for adults with CP (used in Chapter 6). Thirdly, we suggest that it would be very useful to develop or adapt a longitudinal assessment tool to test gross motor function, e.g. the Berman assessment tool (Chapter 4) or the Gross Motor Function Measure (GMFM).

### **Contextual Factors**

In our two projects the main focus was on the 2 components of the ICF-model *Body Function & Structure*, and *Activity and Participation*. We described limited information about the contextual factors, including personal and environmental factors (Figure 7.1). As suggested in some short-term follow-up studies (e.g. age at time of SDR is related to number of orthopaedic surgeries (O'Brien *et al.*, 2004)), it would be very interesting to test if there are any personal or environmental factors which are statistically related, that could perhaps predict outcomes of SDR in the long-term and could be taken into account for patients' selection criteria.

### **International Conformity**

Our projects were conducted in the Republic of South Africa. South Africa is classified by the United Nations as one of the emerging and developing economies. However, it is still a developing country, and as observed by Levin (Levin, 2006), it is important to consider studies on cerebral palsy that are context-specific. Health care (e.g. insurance), environment (e.g. quality of pavements) and culture (e.g. coping) are different in South Africa compared to Western countries. Although it might be methodologically difficult to accomplish, it would be very useful to compare outcomes of different patients living in different cultures and continents.

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