Orthopedic Surgery to Improve Gait in Cerebral Palsy

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2021

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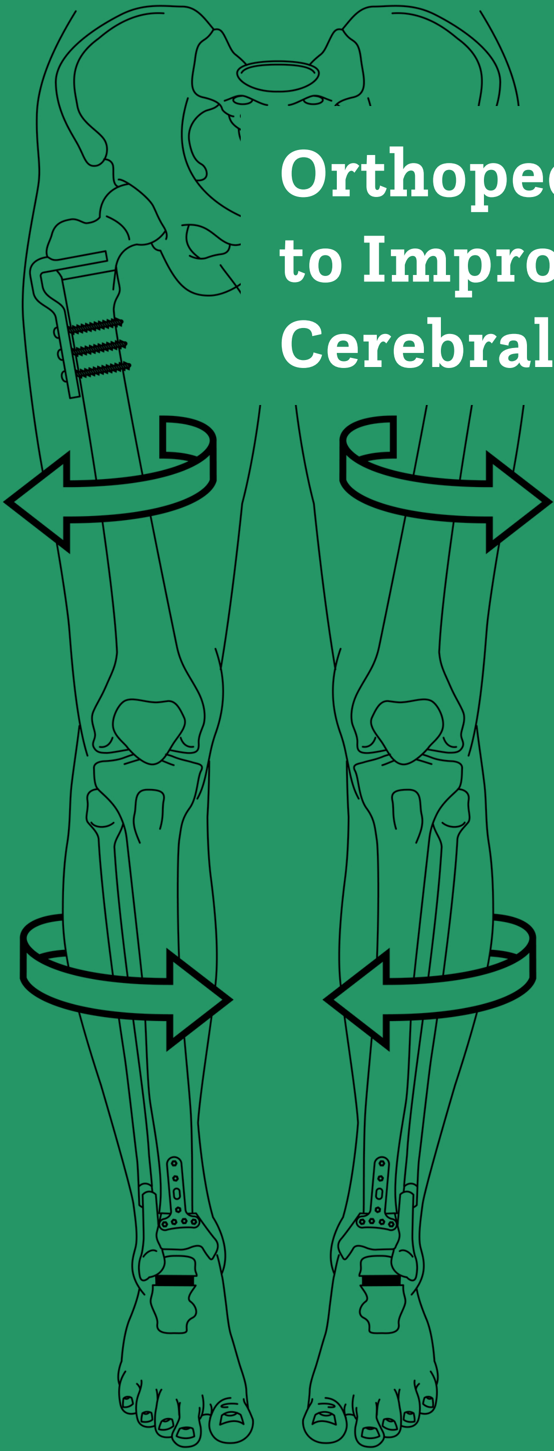

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VRIJE UNIVERSITEIT

Orthopedic Surgery to Improve Gait in Cerebral Palsy

ACADEMISCH PROEFSCHRIFT

ter verkrijging van de graad Doctor of Philosophy aan
de Vrije Universiteit Amsterdam,
op gezag van de rector magnificus
prof.dr. V. Subramaniam,
in het openbaar te verdedigen
ten overstaan van de promotiecommissie
van de Faculteit der Gedrags- en Bewegingswetenschappen
op woensdag 11 juni 2021 om 9.45 uur
in de online bijeenkomst van de universiteit,
De Boelelaan 1105

door

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Abbreviations

3-DGA: 3d gait analysis (same as IGA)

AFO: ankle foot orthosis

BSCP: bilateral spastic cerebral palsy
BoNT-A or BTX: Botulinum toxin type A

CP: cerebral palsy

DF(E)O: distal femoral (extension) osteotomy

ECM: extracellular matrix

FMS: Functional Mobility Scale
FNA: femoral neck anteversion

GGI: Gillette Gait Index
GMFCS: gross motor function classification system
GMFM (-66): Gross Motor Function Measure
GPS: gait profile score
GRF: ground reaction force
GVS: Gait Variable Scores

HRQoL: health related quality of life

ICF-CY: International Classification of Functioning, disability and health for Children and Youth
IGA: instrumented gait analysis (3-DGA for 3d gait analysis)
ITD: Intrathecal baclofen (via pump)

LCP: locking compression plate

MAP: movment analysis profile
MCID: minimally clinical important difference (of GPS)
MTU: muscle-tendon-units

PCSA: physiological cross-sectional-area
PODCI: Pediatric Orthopaedic Data Collection Instrument

RCH: Royal Children's Hospital Melbourne, Australia
RCT: randomized controlled trial
ROM: range of motion

SD: standard deviation
SDR: selective dorsal rhizotomy
sEMG: surface Electromyography
SEMLS: single-event multilevel surgery
ST: semitendinosus muscle

TAL: tendo Achilles lengthening
TATS: tibialis anterior tendon shorthening

UKBB: Universitätskinderspital Beider Basel, Switzerland

WHO: World Health Organization

Chapter 1

General Introduction

Cerebral palsy (CP) is the most common cause of childhood-onset, lifelong physical disability, with an estimated prevalence of 17 million people worldwide. CP is not a disease entity in the traditional sense, but a clinical description of children, who share features of a non-progressive brain injury or lesion acquired during the antenatal, perinatal or early postnatal period till the first birthday.

The clinical manifestations of CP vary greatly in the type of movement disorder, the degree of functional ability and limitation and the affected parts of the body. There is currently no cure, but progress is being made in both the prevention and the treatment of functional problems such as gait improvements.

Clinical management of children with CP is directed towards maximizing body function, activities and participation and in contrast minimizing the effects of the factors that can make the condition worse, such as progressive deformities and contractures of the musculoskeletal system, epilepsy, feeding problems, hip dislocation and scoliosis. These management strategies include: enhancing neurological function during early development; managing medical co-morbidities, muscle weakness and hypertonia; using rehabilitation technologies to enhance motor function; and preventing secondary musculoskeletal problems.

From birth on, many different treatment modalities are needed to guide growth and development of the child, such as physioherapy, orthotic treatment, spasmolytic medication and orthopedic surgery. Very often, bony deformations occur and the orthopedic treatment should start at a very early time point. At the beginning of these treatments conservative options such as physiotherapy and orthotics are dominant, but at older age the role of orthopedic surgeries becomes more and more important.

Orthopedic surgery of bones and soft tissue occurs when the range of joint motion is severely impaired and is considered an important tool to manage developed musculoskeletal deformities. Gait improvements are realized by single level surgery or single-event mulitilevel surgery (SEMLS). However, although in the short term mostly beneficial, in the longer term during further growth, on going non-surgical treatment will still be necassary to prevent relaps. In some cases repeated surgery is indicated. Improvement of surgery in treatment of children with CP requires a profound insight in effectiveness and mechanisms of the different surgical approaches.

1.1. Introduction and definition of cerebral palsy

The term cerebral palsy (CP) does not refer to a specific disease entity, but describes a group of conditions with variable severity (1). CP is a convenient term to describe pathological neurodevelopmental conditions, which are recognized in childhood and persist throughout life. The 2005 International Committee definition is as follows (1, 2):

> *„Cerebral palsy describes a group of disorders of the development of movement and posture, causing activity limitation that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain.*
>
> *The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, cognition, communication, perception, and/or behaviour, by epilepsy and by secondary musculoskeletal problems"*

CP is the most common cause of physical disability affecting children in developed countries, with a prevalence of about 2.0 per 1000 live birth. CP can be the result of a malformation, injury, or infection of the developing brain in utero, during birth, or in very early childhood before the first birthday.

The majority of causes have antenatal antecedents, and many causes are multifactorial. A minority are result of birth trauma or asphyxia. Prematurity and low-birth-weight-for-gestational-age, are the leading associations with CP in developed countries. About 20% of CP has an unknown aetiology. However, an increasing number of genes are discovered, related with the presence of a bilateral spastic paresis. In these cases, no abnormalities on MRT-brain scans are present.

1.2. Classifications of cerebral palsy

CP may be classified by *motor disorder type*. Motor type is classified using terms to describe muscle tone and abnormal movements (SCPE reference manual; (3)):

- **Spastic**: ca. 70%, characterized by increased muscular tone, increased reflexes, pathological reflexes like Babinski-reflex resulting in abnormal pattern of movement and posture.
- **Dyskinetic**: ca. 25%, characterized by involuntary, uncontrolled, recurring, occasionally stereotyped movements, primitive reflex patterns dominate, muscle tone is varying.

- **Ataxic**: ca. 2.5%, characterized by loss of orderly muscular coordination so that movements are performed with abnormal force, rhythm and accuracy.
- **Mixed types:** ca. 2.5%

For clinicians it is important to classify CP also by a topographical distribution: unilateral is involvement on one side of the body, bilateral is involvement on both sides of the body. Bilateral involvement can be subdivided in diplegia, in which the lower limbs are only or more involved than the upper extremity, and tetraplegia, in which the upper extremity are equally or more involved than the lower extremities. The sub-classifications turned out to be not very reliable.

The ICF-CY model

Cerebral Palsy is a life-long condition. To describe the consequences of a chronic medical condition, the World Health Organization (WHO) developed the International Classification of Functioning, disability and health for Children and Youth, the ICF-CY (http://apps.who.int/iris/bitstream/10665/43737/1/9789241547321_eng.pdf).

The ICF-CY model describes the consequences of a decrease on the level of body functions and structures, activities and participation (Fig. 1). External and personal factors modify the effects on each level.

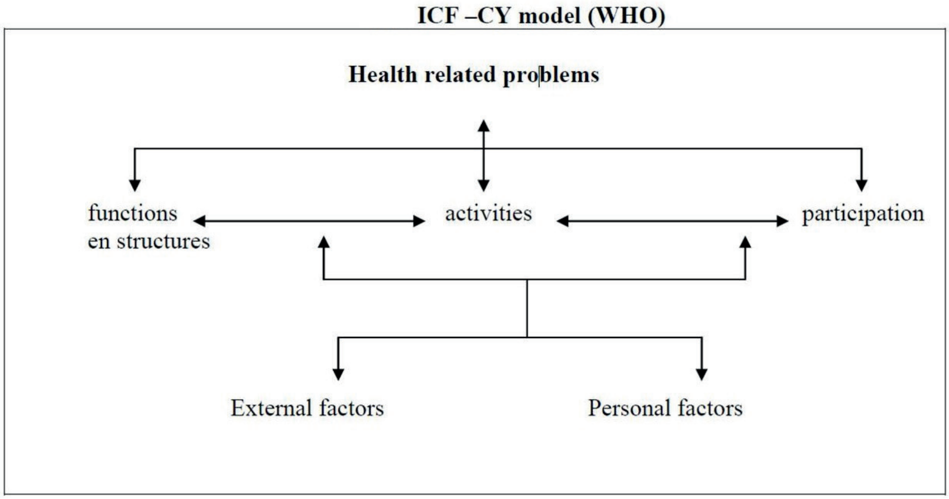

Fig. 1. Description of the ICF-CY model. This model describes the consequences of a decrease on the level of body functions and structures, activities and participation. External and personal factors modify the effects on each level.

In Cerebral palsy, these consequences are described on the level of body function and structures. These include muscle weakness and muscle spasticity, but also cognitive impairments and visual impairments. Therefore, the consequences for daily life are described as the consequences on the level of activities and participation.

Mobility

Orthopedic surgical treatments are focussed on the level of impairments of body function, like decrease of passive range of motion (PROM), muscle shortening, bony deformities, to improve activities in the domain mobility (i.e. lying, sitting, standing, and mainly walking).

The limitations in mobility (according ICF-CY) can be classified with the Gross Motor Function Classification System (GMFCS) (4). The GMFCS is a 5 level classification system that describes the gross motor function of children and youth with cerebral palsy on the basis of their level of activities with particular emphasis on sitting, walking, and wheeled mobility. From the age of 6 years, distinctions between levels are based on abilities, the need for assistive technology, including hand-held mobility devices (walkers, crutches, or canes) or wheeled mobility, and to a much lesser extent, quality of movement. The focus of the GMFCS is on determining, which level best represents the child's or youth's present abilities and limitations in gross motor function (ICF-CY domain mobility). Emphasis is on usual performance in home, school, and community settings focusing on what they really do, rather than what they are able to do at their best (capability). It is therefore important to classify current performance in gross motor function and not to include judgements about the quality of movement or prognosis for improvemen (Fig. 2).

Depending on the social circumstances, walking is to some extend related with participation. Gait parameters are considered to be on the level of body function, only walking distance and walking speed are on the level of activities.

Classification of gait patterns

As orthopedic surgery is mainly focussed on improvement of walking ability, the classification of gait patterns may offer a framework to describe intervention. Several classifications of gait patterns in CP have been described (5).

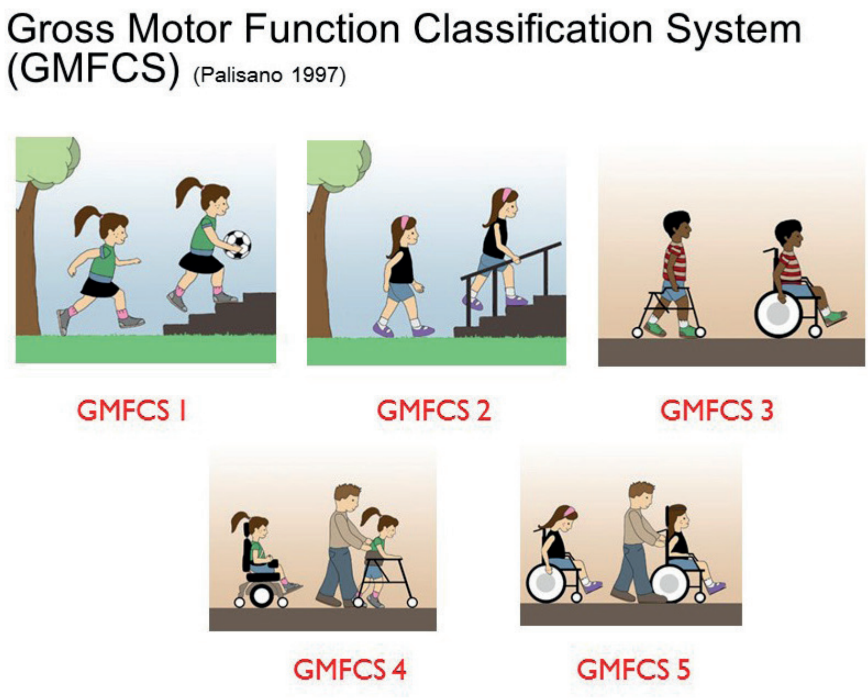

Fig. 2. Classification of gross motor function for children from the age of 6 years (domain mobility of ICF-CY). GMFCS I: children with only limitations in jumping and running; GMFCS II: children who can walk without walking aids, limited in walking distance and need a bannister for climbing stairs; GMFCS III: children who can walk outside only with walking aids; GMFCS IV: children who can move around in a (powered) wheelchair; GMFCS V: children can not move around independently, only with very special equipment.

Gait classification for Unilateral Spastic Cerebral Palsy

In 1987 Winters, Gage and Hicks classified the sagittal gait patterns in unilateral spastic cerebral palsy (Unilateral Spastic Cerebral Palsy or spastic hemiplegia) in a cross-sectional study, based on three dimensional (3D) kinematics (1).

Type I Hemiplegia

In type I hemiplegia, there is a foot drop in the swing phase of gait on the affected side. This is due to loss of selective motor control or weakness in tibialis anterior. There is no contracture of the gastrocsoleus and second rocker in gait is relatively normal.

Type II Hemiplegia

In type II hemiplegia, there is foot drop in swing phase but there is also equinus during stance phase. There is spasticity in the gastrocsoleus, which gradually becomes fixed resulting in an equinus contracture. There can also be equinovarus or less commonly, equinovalgus.

Type III Hemiplegia

In type III hemiplegia, involvement moves proximally to include the knee as well as the ankle level. There is equinus at the ankle and a reduced range of motion at the knee with co-contraction of the hamstrings and rectus femoris. This sub-type is not common.

Type IV Hemiplegia

In type IV hemiplegia, involvement moves proximally to include the hip, as well as the knee and ankle. It also extends from the sagittal plane into the coronal and transverse planes:

There is equinus at the ankle, a flexed, stiff knee, and involvement at the hip, with incomplete hip extension. In the coronal plane there is excessive hip adduction and in the transverse plane, excessive internal rotation and ipsilateral pelvic retraction. Contractures are present at the ankle (equinus), knee (flexion deformity) and hip (adduction and flexion). There is usually increased femoral neck anteversion (FNA) on the affected side, which contributes to hip dysplasia (for details see chapter 5).

Gait classification for Bilateral Spastic Cerebral Palsy

Rodda and Graham extended the Winters, Gage and Hicks classification to include the coronal and transverse planes and made it applicable also for the bilateral spastic CP (BSCP). In their prespective or review Rodda and Graham described four main types of gait patterns for BSCP children (6):

True Equinus

True equinus is characterised by the child walking in equinus in whole stance phase with the knees and hips extended. It is commonly seen in younger children with BSCP when they first learn to walk. The plantar-flexion knee-extension-couple is overactive and the ground reaction force (GRF) is directed in front of the knee throughout the stance phase of gait.

Jump Gait

Jump gait is characterised by equinus in whole stance phase associated with incomplete extension at the knee and hip in late-stance. There is usually excessive knee flexion at initial contact with rapid extension in later stance to a near normal range. In other children, there is more severe knee flexion throughout stance, combined with incomplete hip extension. This is the most common pattern in the pre-adolescent.

Apparent Equinus

Apparent equinus is characterised by toe walking but with near normal ankle range of motion. There is flexion present at the knees and hips. The recognition of "apparent equinus" in contradistinction to "true equinus" is very important to avoid inappropriate lengthening and weakening of the m. gastrocnemius and soleus. Instrumented gait analysis (IGA or 3-D GA for 3D gait analysis) is very helpful in differentiating "apparent equinus" from "true equinus". The "apparent equinus" pattern is often transitional. With further growth and progression of lever arm deformities, the majority of children will develop "crouch gait" (7).

Crouch Gait

Crouch gait is characteried by excessive knee flexion in stance phase, increased flexion at the hip and excessive ankle dorsiflexion (pes calcaneus). Knee stiffness in swing is common. The m. soleus is excessively long and usually weak. This is a very common gait pattern in adolescence and may be part of the natural history of bilateral spastic CP, but is more often caused by isolated tendo Achilles lengthening (TALs) (7, 8). A key feature of crouch gait is that the majority of muscle-tendon-units (MTU) are excessively long. This is by definition true for all of the one-joint muscles such as soleus, vasti and gluteus maximus, and often for the two-joint hamstring muscles. The only consistent contractures are of the m. iliopsoas. In crouch gait, the hamstring muscles are short and/or stiff only in patients with a posterior pelvic tilt. When the pelvis is in the neutral range, the hamstring muscles are of normal length and when the pelvis is anteriorly tilted, the hamstring muscles are likely excessively long. The popliteal angle is decreased while the hamstrings feel tight and are often incorrectly assumed to be short (7).

Diagnostics of gait

Gait analysis is widely used to study the gait pattern. There are different techniques of describing gait:

- Structured observation (as the EVGS, Edinburgh Visual Gait Scale) (9)
- (bi-planar) video observation (9)
- 3-D gait analysis (3-DGA) with active and passive marker systems (1, 10, 11)

The video observation and 3-D gait analysis can be combined with registration of the ground reaction forces and surface-Electro-Myography (sEMG).

At all 3 anatomical levels of the lower extremity (foot & ankle, knee, hip joint and pelvis) limitations in ROM may occur. To treat these limitations adequately, children need treatment at different muscles and/or joints. In patients with complex gait deviations, 3-DGA is usually performed to determine optimal therapeutic strategies. With this method pelvis, hip, knee, ankle and foot joint angles are measured in 3 planes (sagittal, coronal, and transveral) at all three anatomical levels (i.e. foot & ankle, knee, and hip joint).

In addition to 3-DGA, data of the physical examination have to be present for the interpretation of the gait abnormalities. Limited passive range of motions (PROM) at three different anatomic levels (foot & ankle, knee, and hip) during gait are the main problem for gait abnormalities in spastic CP. Limited PROM is always a secondary problem due to abnormal muscle function and different motor type disorders such as spastic, dyskinetic, ataxic or mixed gait pattern. In addition, pain may deteriorate or aggravate gait problems. Pain is determined and measured by a likert scale from 1 to 10, usually.

The *natural history of gait* in a subgroup of children with bilateral spastic CP is one of *deterioration*. In a longitudinal cohort study in children aged 7-13 years, the level of mobility was stable. However, children with a low level of selectivity in voluntary movement ability in the lower limbs showed deterioration in the course of time (12).

Therefore, without intervention gait and function in these children may deteriorate in time, particularly during the pubertal growth spurt (13, 14). In crouch gait, there is also excessive hip flexion, although the pelvic position is variable and can be posterior, neutral, or anteriorly tilted (15).

Of all diagnostic techniques to assess gait, the 3-DGA is most informative as this provides insight in deviations at different joint angles which are measured in three planes (sagittal, coronal, and transveral).

1.3. Treatment

Treatment strategies can consist of a variety of options such as:
- orthopedic surgery,
- physical therapy,
- pharmaco-therapeutic treatment (e.g. Botulinum toxin type A (BoNT-A or BTX-A), Intra-thecal Baclofen (ITB)),
- neurosurgical procedures (Selective Dorsal Rhizotomy (SDR), Deep Brain stimulation), or
- orthotics.

For orthopedic surgical treatment there are many possibilities, but generally it includes corrections of bony deformities and soft tissue procedures. Most of the pediatric orthopedic gait studies report on results of surgical gait corrections, but conservative treatments such as physiotherapy, occupational therapy and orthotic management may play an important role as well. So called “surveillance programs” as used in Sweden and Australia and consisting of clearly defined intervals of clinical and radiological follow-up of a child with CP, become more and more important (16).

Often several surgical interventions are performed in one session, i.e. single-event multilevel surgery approach (17) (SEMLS). Therefore, SEMLS is defined as at least four surgical procedures, performed on two different anatomic levels (hip, knee, or ankle) and on both sides of the body (Fig. 3). The surgical procedure does not need to be symmetrical, but individually customised to the child’s needs.

SEMLS is performed in order to prevent deterioration of gait in patients with bilateral involvement of the lower extremities. This multilevel approach was first described in the 1980’s (18-20). This approach reduces the amount of hospital stays for the child (every year a surgery is known as the so called “birthday syndrome”) and corrects all deformities in one major operative session (1, 2). Usually this approach is used in children with GMFCS levels II and III.

Very little is known about how the surgical interventions improve gait compared with when surgical intervention was withhold. Effects of surgical treatment on gait characteristics in children with BSCP have been mostly investigated by follow-up measurements, but without a no-treatment control group (21). To the best of our knowledge the study by Bell et al.(14) is the only study in which stride parameters and kinematics over time were assessed in children with BSCP referred for advice for surgical treatment.(21) This study clearly shows a deterioration of gait, particularly at the level of the ankle and knee joints. There was no surgical intervention in this cohort during the study period of 4.4 years. Of the 28, 19 children were classified with the diagnosis of BSCP, 7 with unilateral CP, and 2 with total body involvement.

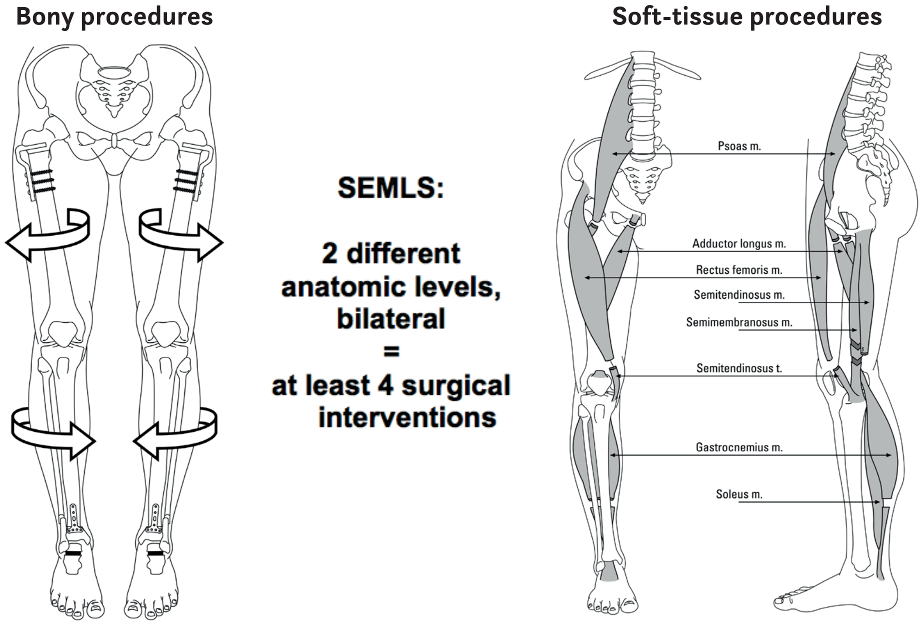

Fig. 3. On the left side, common bony procedures are represented: femur exorotation osteotomy, tibia endorotation osteotomy, triple arthrodesis of the ankle. On the right side, muscle procedures are represented: psoas tenotomy, adductor tenotomy, rectus femoris tenotomy, hamstrings elongation, gastrocnemius and soleus elongation.

In general, SEMLS results in clinically significant improvements in gait and function, in children with BSCP, which were maintained at 5 years after surgery (22).

Severe gait dysfunction in patients with spastic diplegia has been reported (2) to be improved 5 years after the index surgery in one operative session by single-event multilevel surgery. In our study (2) additional surgery after the index procedure was performed in 9 of the 14 patients, because relapse of the original or new gait problems had occured (see chapter 8). In summary, in children with CP severe gait dysfunction can be corrected effectively in the short term by using the SEMLS approach, but there is currently no cure. Clinical management of children with CP is therefore directed towards maximizing body function, physical activities and societal participation. Because there is no cure to treat the underlying neurological problems, there is a risk for relapse after SEMLS. The results of the Swedish national CP register with systematic surveillance and treatment reported promising results: complete prevention of hip luxation, reduction of the need for surgery to 1/3 in comparison with the numbers before the start of the register (23).

Although over the years improvements have been made, there is still place for improvement of the SEMLS (24, 25). To this end a more detailed insight into the mechanisms and effectiveness of SEMLS is required.

In spastic CP a limited joint range of motion (ROM) during gait is primary caused by the neurological impairments, but during growth secondary bone and muscle tissue show alterations in their development and these also may contribute to limited ROM.

These mechanisms underlying impaired gait, relate to an enhanced muscle activity, increased stiffness and/or shortening of muscle and tendon structures as well as to joint and bone problems.

The rationale for orthopedic surgery (single level or SEMLS) is the improvement of ROM at least one but generally at different levels (foot & ankle, knee, hip) during gait and therefore to improve gait dysfunction.

> *Treatment principles* must be individually selected to the child's needs. Questions as which problem on the level of body function and activity has to be solved, and what are the expectations after surgery on physical activity, as determined by a comprehensive evaluation need to be addressed. This includes careful physical examination by radiological analysis and instrumented gait analysis (3-DGA). The aims of orthopedic treatment should be pain reduction or no pain at all, and improvement in the domain mobility (walking, as well sitting, standing and transfer).

When limitations in passive range of motion by muscle shortening and/or bony deformity have developed, orthopaedic surgery is indicated to improve body functions.

This overall aim of this thesis is to obtain a retrospective overview of how successful orthopedic surgical procedures are regarding the correction of overall gait dysfunction and to investigate the effectiveness of SEMLS, and of particular surgical interventions at single joint levels.

1.4. Outline of the thesis

The aim of this thesis was to obtain insight in how different single-level or single-event multilevel surgery (SEMLS) procedures of the lower limb in children with Bilateral Spastic Cerebral Palsy affect gait.

For this purpose we have investigated results of surgical interventions at three levels i.e. the foot & ankle joint, the knee joint, and the hip joint.

Limited range of motion (ROM) at these three different anatomic levels (foot & ankle, knee, and hip) during gait is the main problem for gait abnormalities in spastic CP. In addition pain may deteriorate or aggravate gait problems in children with CP.

Orthopedic surgery at a single joint/level or at multiple levels is directed at the management of orthopedic problems, such as:

- Limited joint ROM and pain, and
- Improvement of patient's level of mobility and activity.

Therefore depending on where the orthopedic problems are located, single or multilevel surgery is performed to improve gait dysfunction and gait deviations. The indications for these treatment options are different:

1. Single level surgery: addresses just one orthopedic problem at one anatomic level (e.g.: equines at the level of foot & ankle)
2. Single-event multilevel surgery (SEMLS): contains a minimum of 4 different surgical procedures (each 2 left and right) at 2 different anatomical levels. The interventions could either be bony or soft-tissue procedures.

Kinematic changes are described by using Gait Profile Score (GPS) and Movement Analysis Profile (MAP) as described by Baker et al. (10, 26), however very little is known about the variability improvements in gait quality, stability and durability of these results over decades.

The essential questions are "does orthopedic surgery in cerebral palsy improve":

- the level of physical activity?, and
- specific the level of mobility?, and
- are ROM and gait kinematics improved?, or
- how long the effects will last?

Data for studies discussed in chapters 2, 3, 4, 6, and 8 are from the University Children's Hospital Basle, UKBB, Switzerland. Data for studies discussed in chapters 5, 7, and 9 are from the Royal Children's Hospital, RCH, Australia.

In chapter 2 we tested whether a preoperative botulinum toxin (BTX-A) test injection is able to filter out patients who are at risk for deterioration after muscle-tendon lengthening surgery?

In chapter 3 we investigated whether the combined Tendon Achilles lengthening (TAL) and Tibialis Anterior Tendon Shortening (TATS) improves gait in spastic equinus and droop foot in swing phase?

Chapter 4 reports for the same cohort the long-term outcomes after 5.8 years with respect to the necessity of the use of an ankle foot orthosis (AFO) postoperatively.

We investigated the outcomes of 11 children with spastic hemiplegia Type IV after unilateral multilevel corrections to improve ROM around ankle and hip. The results of this study are presented in chapter 5.

In chapter 6 we present results of a study in which we investigated the influence of distal surgical corrections of bone (extension osteotomy of the distal femur) and soft tissue (patellar tendon shortening) at the level of the knee on hip extension.

In chapters 7,8, and 9 we reported the outcomes of SEMLS using Movement Analysis Profile (MAP) and Gait Profile Score (GPS) (10) on different aspects of outcomes, such as stability of the GMFCS levels (chapter 7), correction of severe crouch gait (chapter 8), and the influence of different factors on the outcomes of SEMLS (chapter 9).

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1

Chapter 2

Preoperative botulinum toxin test injections before muscle lengthening in cerebral palsy

Erich Rutz, Eva Hofmann, and Reinald Brunner

Abstract

Background. Muscle weakening is a well-known side effect of muscle–tendon lengthening. Botulinum toxin A (BTX-A) weakens the muscle temporarily by blocking the neuromuscular junction.Hence application of the drug is a logical step to test whether weakness deteriorates function prior to an operation. In the present study, BTX-A application is used to test preoperatively whether the gait pattern depends on the strength of the tested muscle. Since 1999, instrumented gait analysis, including kinematic, kinetic, and dynamic electromyographic data, is routinely used to define the individual surgical program.

Methods. In our series of 110 consecutive patients with cerebral palsy (CP) considered for surgical muscle lengthening from 1999 to 2008, BTX-A was applied to identify patients at risk for functional deterioration. Gait analysis was repeated 6 weeks (maximum effect of BTX-A) and 12 weeks (follow-up) after the test injection to check for loss of joint control (excessive ankle dorsiflexion, knee flexion, increased anterior pelvic tilt).

Results. In all, 20.9% (*n* = 23) showed deterioration in gait after preoperative BTX-A test injections (*n* = 112, two patients had two test trials) in all muscles considered for lengthening. As a consequence, their lengthening surgery was canceled. A total of 68 patients underwent surgery as planned, and in none of them did gait function deteriorate. These clinical data were compared to those of a historical group (*n* = 105) before this test, where 18% showed functional deterioration after surgery. The similar percentage of patients filtered out by the test suggests that there could be a context to the number of poor results in the historical group.

Conclusions. We conclude that preoperative BTX-A test injection is a reliable tool for filtering out patients with risk of deterioration after muscle lengthening surgery in patients with CP and can be helpful to avoid poor outcomes.

Introduction

Botulinum toxin A (BTX-A) blocks the release of acetylcholine at the neuromuscular junction, which partially denervates the muscle and thus decreases muscle strength and tone. Intramuscular BTX-A injection is a local reversible treatment with a wide range of therapeutic applications, including temporary reduction of spasticity. BTX-A injections in children with cerebral palsy (CP) have been shown to be safe with relatively few side effects. Today it is accepted as a standard antispastic therapy.[1,2]

Muscle–tendon lengthening surgery always carries the danger of muscle weakness.[3–5] Overlength and consecutive muscle weakness should be avoided, especially as repairs of failed muscle–tendon lengthening are difficult or even impossible. In contrast, correctly indicated lengthening maintains muscle force and provides good clinical results in the long term.[5,6] Most of lengthening is performed intramuscularly, although tendon lengthening is more powerful for gaining length but bears a higher danger of postoperative muscle weakness.[7–9] According to two recently published cohortstudies,[10,11] crouch gait results in 42%–60% of the patients after Achilles tendon lengthening. De Morais Filho et al.[10] showed that the mean anterior pelvic tilt changed from 12° to 22° after medial hamstring lengthening. Hence, patients with CP should be selected carefully for muscle lengthening to avoid a poor surgical outcome or even loss of walking ability.

Administration of BTX-A weakens the muscle, with its maximum effect nearly 6 weeks following the initial injection. This effect is temporary, lasting approximately 12–16 weeks. It may be used to test whether the patient's function depends on the strength of a specific muscle. Even in structural alterations (e.g., fixed equinus or fixed flexion contracture of the knee) whose correction cannot be simulated by intramuscular BTX-A injections, additional weakening can still result in deterioration. In such cases, surgery with a similar effect would be deleterious.

We retrospectively compared the functional outcome of a cohort before the test period with a cohort of tested patients. The aim of the present study was to investigate the hypothesis of functionally testing the effect of muscle weakening by BTX-A test injections to avoid poor outcomes of surgical muscle–tendon lengthening in patients with CP.

Patients and methods

Patients

A retrospective analysis was performed that included all consecutive CP patients during 1999–2008 who were given single or multilevel BTX-A injections fortesting the possibility of muscle lengthening.

Methods

All patients underwent a first session of instrumented gait analysis, including kinematics, kinetics, and dynamic electromyographic (EMG) data, using a motion capture system (six camera VICON 460 system; Oxford Metrics, Oxford, UK), two force plates (Kistler Instrumente, Winterthur, Switzerland), and an eight-channel surface EMG system (Zebris, Tübingen, Germany). The patients walked at their self-selected speed. The Helen Hayes Marker set[12] was used, and at least six trials were recorded. Anthropometric data were recorded for appropriate scaling. Surface EMG was recorded simultaneously. Bipolar Ag/AgCl surface electrode pairs (electrode diameter 10 mm, interelectrode spacing 22 mm) were placed bilaterally over the medial gastrocnemius, tibialis anterior, rectus femoris, and semitendinosus muscles. For electrode placement, the SENIAM[13] recommendations for surface EMG were followed. The ground electrode was placed over the tibial tuberosity. The EMG signals were bandpass filtered (10–700 Hz) and collected at a sampling rate of 2500 Hz. All data were expressed as a percentage of the gait cycle using Polygon software (Oxford Metrics).

The surgical procedure was planned on the basis of this analysis and could include complex multilevel approaches as well as single muscle lengthening. BTX-A was next injected under ultrasonography (US) control in all muscles considered for lengthening. The average dose was 50 U of BTX-A per muscle belly (range 25–100 U Botox). The injection was performed according to the European consensus table on botulinum toxin for children with CP.[14]

Gait analysis was repeated 6 and 12 weeks after the BTX-A injection to check for the shortand mid-term effect of muscle weakness. Excessive ankle dorsiflexion at the terminal stance of the gait cycle, loss of knee extension at the mid and terminal stance, and/or increased anterior pelvic tilt during the whole gait cycle showed a loss of joint control and hence were considered deterioration. The first two criteria describe crouch gait, and the last point is a well-known adverse effect of hamstring overlengthening. Hyperextension of the knee joint at the terminal stance of the gait cycle was not defined as deterioration. The gait cycle, which best represented the kinematic pattern, was selected from six trials by a human movement scientist or a physiotherapist trained in gait analysis. In cases with high variability among the six trials, all of the files were carefully reviewed by one of two of the authors (E.R., R.B.) The diagnosis of functional deterioration was based on this selected gait cycle; and, consequently, surgery addressing the respective muscle groups was omitted.

In the BTX-A test cohort, a total of 110 patients had 112 test trials (two patients had two test trials of BTX-A): 67 boys and 43 girls (mean age 14.4 years; median 17 years; range 8–53 years). A total of 42 patients suffered from hemiplegia, 39 spastic diplegia, 27 quadriplegia, and 2 ataxic CP. Altogether, 28 patients had Gross Motor Function Classification System (GMFCS) level I, 76 patients had GMFCS level II, and 6 patients had GMFCS level III. No patients with GMFCS level IV or V were included. In all, 23 patients were rejected from surgery owing to the test result, 9 refused surgery, and 10 have surgery planned but it has not yet been done. The clinical outcome of the remaining 68 patients was compared to a historical cohort (1990–1998, $n = 105$) without such a BTX-A test injection.

In the historical cohort, not all of the 105 patients underwent instrumented gait analysis, as this assessment was not routinely done even for minor muscle problems at that time. Poor results in the historical cohort were defined as: dorsiflexion of the ankle >20° during the whole gait cycle, knee flexion >30° at the terminal stance phase, hyperlordosis of the lumbar spine during the whole gait cycle (as a sign of loss of pelvic control), or poor result (identical definitions as for the tested cohort) in gait analysis after 1999. These criteria were evaluated during the most recent clinical followup by visual gait analysis. The GMFCS levels for all patients were retrospectively classified by scanning all the surgical reports and patient's histories by the first author. In all, 17 patients had GMFCS level I, 84 patients had GMFCS level II, and four patients had GMFCS level III. No patients with GMFCS level IV or V were included in this study. Statistically, there was no difference between the two cohorts concerning the general parameters ($P = 0.17$) of age, sex, and diagnosis (Table 1).

All patients/parents gave written consent for research purposes in accordance with local ethics committee requirements. The study was performed according to the declaration of Helsinki (World Medical Association).

Table 1. Comparison of historical and current cohorts (before surgical intervention).

Parameter	Historical cohort (without BTX test)	Current cohort (with BTX test)
Study period	1990–1998 (9 years)	1999–2008 (10 years)
No. of patients (n)	105	110
Patient status		
Hemiplegic	44 (41.9%)	42 (38.2%)
Diplegic	37 (35.2%)	39 (35.5%)
Quadriplegic	23 (21.9%)	27 (24.5%)
Ataxic	1 (1.0%)	2 (1.8%)
GMFCS levels (no. of patients)		
I	17 (16.2%)	28 (25.5%)
II	84 (80.0%)	76 (69.1%)
III	4 (3.8%)	6 (5.4%)
Age (years)	14.1 (7–49)	14.4 (8–53)
Sex (F/M)	42/63 (40.0%/60.0%)	43/67 (39.1%/60.9%)
No. of BTX tests	0	112 (two patients with two trials)

BTX, botulinum toxin

Statistics

Data were assessed using Student's *t*-test (SPSS software, version 15.0; SSPS, Chicago, IL, USA). The significance level was set at $\alpha = 0.05$. A chi-squared test was used to compare the clinical outcome. A power analysis was not carried out because of the limited numbers of patients in each cohort.

Results

A total of 23 patients (20.9%) (8 hemiplegia, 8 quadriplegia, 6 spastic diplegia, 1 atactic CP) showed deterioration according to the gait analysis 6 weeks after the BTX-A test injection. Five of these patients had BTX-A test injections in the hamstrings and the gastrocnemii, seven in the hamstrings only, five in the gastrocnemii only, four in all heads of the triceps surae, and two in other muscles (one in the tibialis anterior muscle and one in the tibalis anterior, tibialis posterior, and rectus femoris muscle). The gait analysis 12 weeks after the test injection showed stabilization or even improvement of the gait patterns in all of the patients as the effect of BTX-A faded. Figures 1 and 2 show two examples of deterioration after the BTX-A test injection.

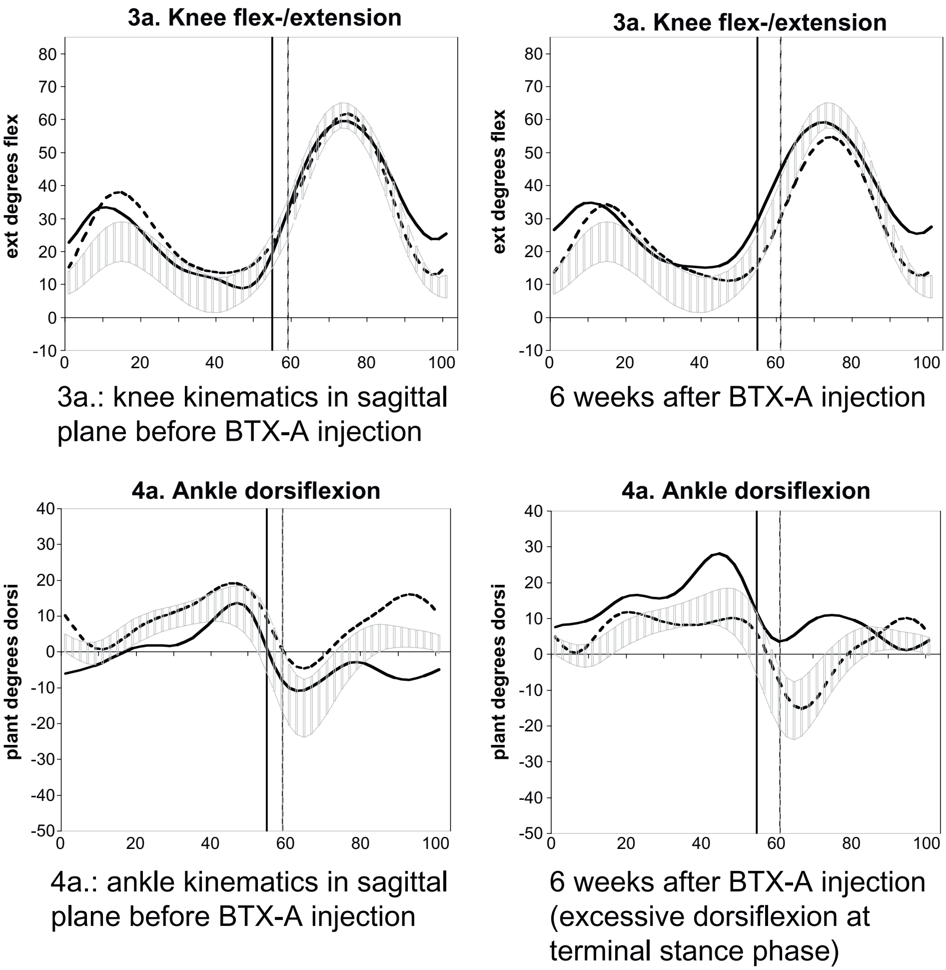

Fig. 1. Deterioration of gait pattern in a 11.5 years old left hemiplegic girl 6 weeks after botulinum toxin-A (BTX-A) test injections in both heads of the gastrocnemius muscle. The *solid line* represents left side, the *dashed line* right side.

Altogether, 87 patients (79.1%) showed improvement or no deterioration of the gait pattern according to the gait analysis 6 weeks after the BTX-A test injection. The gait analysis 12 weeks after the test injection showed stabilization or even improvement of the gait patterns in all of the patients as the effect of BTX-A faded. Nine patients rejected surgery despite a favorable test result. Ten patients agreed to the proposed operation and await surgery. In all, 68 patients had surgery (38 had Achilles tendon lengthening, 30 had hamstring lengthening), with no postoperative deterioration. In the BTX-A test cohort, no deterioration was found after surgery (Table 2).The mean clinical followup after the surgical intervention was 3.8 years (median 3 years, range 1–9 years). At that time, none of the patients had any change in their GMFCS level.

Thus, the cohort of patients who had single or multilevel BTX-A injections for testing regarding a considered muscle lengthening experienced no adverse effects of the BTX-A.

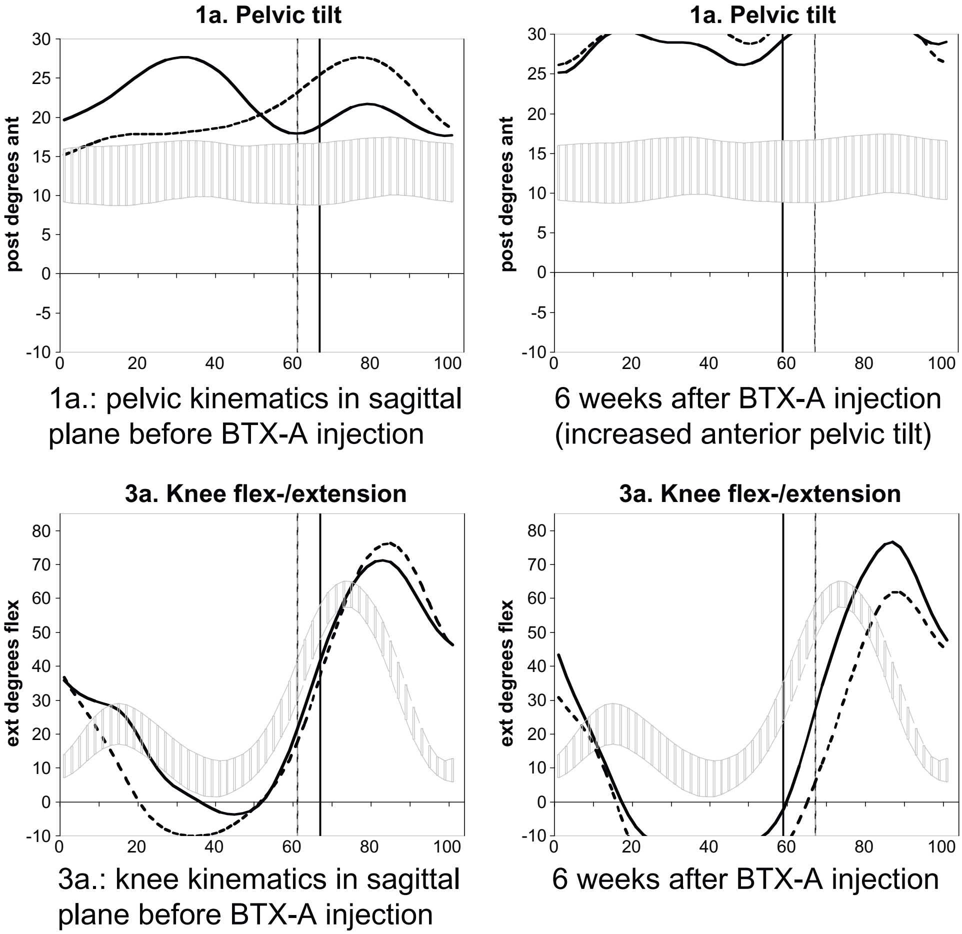

Fig. 2. Deterioration of gait pattern in a 6.5-year-old tetraplegic boy 6 weeks after BTX-A test injections in the bilateral hamstring muscles. The hamstrings are now functionally long. The *solid line* represents left side, the *dashed line* right side.

In the historical cohort of 105 patients, 65 patients underwent Achilles tendon lengthening and 40 underwent hamstring lengthening. In 19 patients (18.1%) there was deterioration after surgery: 12 (11.4%) after hamstring lengthening and 7 (6.7%) after Achilles tendon lengthening. There were more poor results during the early 1990s than during the late 1990s. Comparison of the difference in the clinical outcomes (postoperative results) of the two groups was highly significant ($P = 0.016$) (Table 2). Table 3 shows the postoperative poor results in the historical cohort and their clinical findings. The mean clinical follow-up after the surgical intervention for the historical cohort was 13.6 years (median 13 years, range 10–18 years). At that time, none of the patients had had a change in their GMFCS level.

Table 2. Comparison of historical and current cohorts (after surgical intervention).

Parameter	Historical cohort (without BTX test)	Current cohort (with BTX test)
Period	1990–1998 (9 years)	1999–2008 (10 years)
No. of patients	105	110
Deterioration after BTX test (no.)	Test not available	23 (20.9%) ⟶ no surgical intervention
Improved or same after BTX test (no.)	Test not available	87 (79.1%)
Interventions (no.)	105	68
Postoperative result		
Good	86 (81.9%)	68 (100%)
Poor	19 (18.1%)[a]	0
Patient status		
Waiting for surgery	0	10 (9.1%)
Refused surgery	0	9 (8.2%)

[a] Poor results in the historical cohort: see Table 3

Table 3. Postoperative poor results in historical cohort (1990–1998).

Result	Historical cohort (without BTX test) (no. of patients)
Ankle dorsiflexion >20° (during the whole gait cycle)	7 (36.8%)
Knee flexion >30° (at terminal stance phase)	19 (100%)
Hyperlordosis of the lumbar spine (during the whole gait cycle)	10 (52.6%)
Disappointment with the postoperative result (parents or patients)	19 (100%)
Poor result in gait analysis after 1999	3 (15.8%)

There were 19 (18.1%) patients with poor results in the historical cohort: 7 patients (6.7%) after Achilles tendon lengthening and 12 patients (11.4%) after hamstring lengthening

Discussion

Botulinum toxin-A reliably reduces muscle strength and tone. Today BTX-A injections are accepted as a standard antispastic therapy[1,2,15–19] in children with CP.

The advantageous effect of BTX-A on function is well documented.[18,20–29] The hypothesis of functionally testing the effect of muscle weakening to avoid poor outcomes of surgical muscle–tendon lengthening is tested. This concept is new and, as far as we know, not reported in the literature.

In the present study, BTX-A application was used to test preoperatively whether the gait pattern depends on the strength of the muscle. Injecting BTX-A weakens the muscle, and lengthening (especially of tendons) may have a similar effect. Whereas the effect of BTX-A is reversible, the effect of surgery persists and may be difficult to be repaired in case of failure. We are aware of the fact that only functional problems can be tested with this method and that the effect of a change in structural problems (e.g., a fixed equinus, knee flexion contracture, or bony deformities) cannot be tested by muscle weakening using BTX-A test injections. After canceling all planned surgical lengthening when the test showed deterioration, no poor results occurred.

This study did not include patients with GMFCS level IV or V. Some of these patients, of course, have structural alterations where surgery has to be performed because of pressure points of orthoses or care problems. In all of our patients with GMFCS level I, II, or III in whom there was deterioration after the BTX-A test injections series the surgery was canceled. However, they all had alternative treatments with orthoses (hinged ankle-foot orthoses at the level of the ankle joint and knee-extension splints for at least 1 h per day in patients with short hamstrings). Alternatively, instead of performing Achilles tendon lengthening, a less powerful operation (i.e., the Baumann or Strayer procedure) can be used at the level of the ankle joint if surgery is required. In so doing, the "dose of surgery" could be adapted and a surgical procedure tailored to the needs of the patients is used. In place of semitendinosus tenotomy at the level of the knee joint, only intramuscular lengthening can be done, which is less powerful.

None of the patients had BTX-A injections in the psoas muscle because we are afraid of weakening this muscle too much. In our opinion, the psoas muscle is essential for hip flexion and to climb stairs. Basset et al.[30] showed in 1999 that lengthening the psoas tendon during open reduction of developmental dislocation of the hip is associated with considerable atrophy of the psoas muscle. For this reason, psoas lengthening is, in our view, not a good option; and we prefer extending intertrochanteric osteotomy if indicated.

A weakness of the study might be its retrospective nature with comparison to a historical cohort. This historical cohort did not undergo gait analysis because it was not available in our institution before 1999. It should also be noted that the clinical follow-up time of the two cohorts is not comparable. In our opinion, instrumented gait analysis is more accurate in finding and describing gait deviations. However, the patients do not differ in their general parameters; and the indication for surgical procedures including a multilevel approach has remained unchanged over the last years. Another weakness is the clinical way to assess the recorded gait data: Only one of six trials is chosen and used for the decision making. Nevertheless, as the BTX-A test injection procedure includes this assessing method and the procedure is tested as a whole, this point is, in our opinion, of minor importance.

With the preoperative BTX-A test injections, we were able to identify 21% of patients at risk for functional deterioration in case of muscle weakening. In the historical cohort, 18% showed deterioration after the surgical intervention. The similar percentage of patients filtered out by the BTX-A test injections suggests that there could be a context to the number of poor results in the historical group. For this reason, we conclude that the preoperative BTX-A test injection is a reliable tool for filtering out CP patients who are at risk of deterioration after muscle lengthening surgery and can be helpful for avoiding poor outcomes. As no further deterioration was seen at 12 weeks after the injection, it seems that 6 weeks is sufficient.

Acknowledgments

The authors thank the Swiss National Science Foundation (SNF) for financially supporting this study.

There are no conflicts of interest for any of the authors.

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Chapter 3

Tibialis anterior tendon shortening in combination with Achilles tendon lengthening in spastic equinus in cerebral palsy

Erich Rutz, Richard Baker, Oren Tirosh, Jacqueline Romkes, Celina Haase and Reinald Brunner

Abstract

Equinus is the commonest deformity in cerebral palsy (CP). Many different surgical procedures have been described for the treatment of spastic equinus. In long standing equinus deformities the tibialis anterior muscle becomes elongated which is one reason for muscle weakness. Surgical tendon shortening of the tibialis anterior tendon was therefore introduced to rebalance muscle strength.

All patients with CP who had a tibialis anterior tendon shortening (TATS) in combination with a tendo Achilles lengthening (TAL) were included in this study. A total of 29 patients had 30 surgical interventions (21 hemiplegic patients: 14 boys/7 girls, age 9–22 years; mean 15.2 years; 5 diplegics and 3 quadriplegics; 5 boys/3 girls, age 7–37.5 years; mean 14.8 years). Fifteen patients had additional surgery (soft tissue or bony procedures). The TATS was performed at the distal insertion with transosseous tendon fixation in the medial cuneiform bone at the original place.

Movement Analysis Profile (MAP) for ankle dorsi-/plantarflexion, Gait Profile Score (GPS), Gait Deviation Index (GDI), and Gillette Gait Index (GGI) improved significantly for all patients compared pre- to postoperatively. In 93% of the patients active dorsiflexion of the ankle was possible postoperatively. We conclude that TATS in combination with TAL in spastic equinus in CP is a safe procedure and improves but not completely corrects foot positioning during gait. For the treatment of spastic equinus in CP we recommend shortening of the elongated antagonist (TATS) in combination with lengthening of the short agonist (TAL) for achieving optimal postoperative function.

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Introduction

Equinus is the commonest deformity in cerebral palsy (CP) [1–3] and consequently affects balance, standing, and gait. The treatment plan can consist of a variety of conservative options (physiotherapy, ankle foot orthoses, casting, and injections of botulinum toxin type A) or surgical interventions. Several types of surgical procedures have been described for the treatment of spastic equinus including partial neurectomy of the gastrocnemius [4], lengthening of the origin of the gastrocnemius [5], recession of the proximal gastrocnemius aponeurosis [6,7], aponeurotic lengthening of gastrocnemius and soleus [8] combined with lengthening of the gastrocsoleus fascia [9], lengthening [2] and translocation [10] of the Achilles tendon.

In patients with an equinus deformity walking independently, gastro-soleus muscle-tendon complex lengthening is commonly undertaken to improve gait. Previous studies have reported recurrent equinus in approximately 10–30% of patients with hemiplegia [11,12] and overcorrection with calcaneus foot in 3–30% of patients with spastic diplegia [10,13,14]. In addition Biedermann et al. [15] reported in 2007 a high recurrence rate after calf lengthening with the Ilizarov apparatus for treatment of spastic equinus foot deformity. This technique has a higher recurrence rate than most other operative techniques but carries virtually no risk of overcorrection.

The mildest type of hemiplegic CP solely consists of a drop foot deformity [16]. Whereas central weakness was already suggested by Lin et al. in 1992 [17], imbalance of agonist and antagonist muscle lengths can lead to additional weakness. In case of long standing equinus deformity the antagonist (tibialis anterior muscle) becomes elongated, which weakens the muscle, and the agonist (gastro-soleus muscles) becomes short, which also weakens the muscle. Although dorsiflexion movement increases as a consequence of TAL, additional therapy is required to correct gait. For this rationale, simultaneous tendon shortening of the antagonistic tibialis anterior tendon-muscle unit was introduced to rebalance muscle strength. More complex tendon transfers are used to address the drop foot, such as the ‘‘Bridle’’ procedure [18]. This procedure consists of a tenodesis between the posterior tibialis, anterior tibialis, and peroneus longus, combined with an Achilles tendon lengthening for treating equinus and equinovarus deformities. This technique avoids problems of tendon attachment to bone and tendon placement for balance. To our knowledge the combined surgical technique of shortening the anterior tibialis tendon-muscle unit and lengthening the calf tendon-muscle unit in spastic equinus in CP has not been previously described.

The Movement Analysis Profile (MAP) [19] has recently been developed to summarise much of the information contained within the kinematic data arising from three-dimensional (3D) instrumented gait analysis. The MAP describes the magnitude of deviation of nine individual variables averaged over the gait cycle. The Gait Profile Score (GPS) [19] reduces this information further to give a single number that reflects how much a gait pattern deviates from normal. Hence the GPS is a single index outcome measure that summarises the overall quality of a patient's gait kinematics and is useful for evaluating outcomes [19].

The aim of this study was to evaluate the short-term outcome and the surgical results after tibialis anterior tendon shortening (TATS) in combination with tendon Achilles lengthening (TAL) in spastic equinus in children with CP using the Movement Analysis Profile and the Gait Profile Score. We hypothesised, that the combination of TATS and TAL would correct both the equinus and drop foot deformities.

Materials and methods

A retrospective analysis of all ambulatory children with CP and spastic equinus was performed to evaluate the short-term outcome and the operative results after TATS in combination with TAL. A total of 29 patients had 30 surgical interventions between August 2004 and March 2008. Group I consisted of 21 patients with hemiplegic CP (11 right and 10 left affected) and group II of 5 diplegic (1 bilaterally affected case) and 3 quadriplegic patients. The mean age of all patients was 15.1 ± 6.3 years (median = 15.0 years, range 7.0–37.5 years) at the time of surgery. Nine patients had GMFCS level I, and 20 had GMFCS level II. The mean follow up time for patients in group I was 14.0 months (range: 9.0–24.0) and for patients in group II 14.3 months (range: 9.0–24.0). Active ankle dorsiflexion was not possible in any of the patients preoperatively. Exclusion criteria were a diagnosis other than cerebral palsy, dystonic or mixed movement disorder, Botulinum toxin A injections in the previous 6 months and GMFCS level III or higher.

All patients had pre- and postoperative three-dimensional (3D) instrumented gait analysis undertaken by a physiotherapist and a human movement scientist, both experienced in gait analysis. The clinical assessment included the examination of the passive range of motion (RoM), spasticity according to the modified Ashworth/ Bohannon scale [20] (scale: 0–4), and the manual muscle strength test [21,22] (scale: 0–5) of the ankle dorsi- and plantarflexors. The pre- and postoperative instrumented gait analysis included kinematics, kinetics, and dynamic surface electromyography

(EMG), using a motion capture system (6 camera VICON 460 system, Oxford Metrics Ltd., UK), 2 force plates (Kistler Instrumente AG, Winterthur, Switzerland) and an 8 channel surface EMG system (Neurodata GmbH, Vienna, Austria). Patients walked barefoot at their self-selected speed. The Helen Hayes Marker set [23] was used and at least six trials were recorded. Anthropometric data were recorded for appropriate scaling. Surface EMG was recorded simultaneously. Bipolar Ag/AgCl surface electrode pairs (electrode diameter 10 mm and an inter-electrode spacing 22 mm) were placed bilaterally over the medial gastrocnemius, tibialis anterior, rectus femoris, and semitendinosus muscles. For electrode placement, the SENIAM [24] recommendations for surface EMG were followed. The ground electrode was placed over the tibial tuberosity. The EMG signals were band-pass filtered (10–700 Hz) and collected at a sampling rate of 2500 Hz. All data were expressed as a percentage of gait cycle using the Polygon software (Oxford Metrics Ltd., UK). From the 3D gait data, temporal-spatial parameters (cadence, stride length, and walking speed), the Gillette Gait Index (GGI) [23], the Gait Deviation Index (GDI) [25], the Movement Analysis profile (MAP) [19] and the Gait Profile Score (GPS) [19] were calculated for all patients pre- and postoperatively for group I and II separately.

Surgery

Before TAL a test injection with botulinim toxin type A in the gastro-soleus muscles excluded all patients who deteriorated by weakening this muscle [26]. In all others TAL was performed using an open Z lengthening of the Achilles tendon with the tendon repaired under maximal tension using an absorbable suture (6 Vicryl[1], Ethicon Inc., Johnson and Johnson) and the foot in 108 of dorsiflexion in patients with hemiplegia and 5–108 of plantarflexion in diplegia or quadriplegia.

TATS was performed distally with transosseous fixation to the medial cuneiform bone (suture with 6 Vicryl[1], Ethicon Inc., Johnson and Johnson) at the original insertion. The tendon was spanned as much as possible, so that the foot held itself in a plantigrade position (Fig. 1). All surgical procedures were performed by the first and last authors.

Postoperative treatment

After the surgical procedure a cast was applied in plantigrade (hemiplegia) or mild equinus position (diplegia and quardriplegia) for 6 weeks postoperatively, with weight bearing after 4 weeks. If active ankle dorsiflexion was not possible immediately after cast removal a hinged ankle foot orthosis (AFO) with plantarflexion block was fitted.

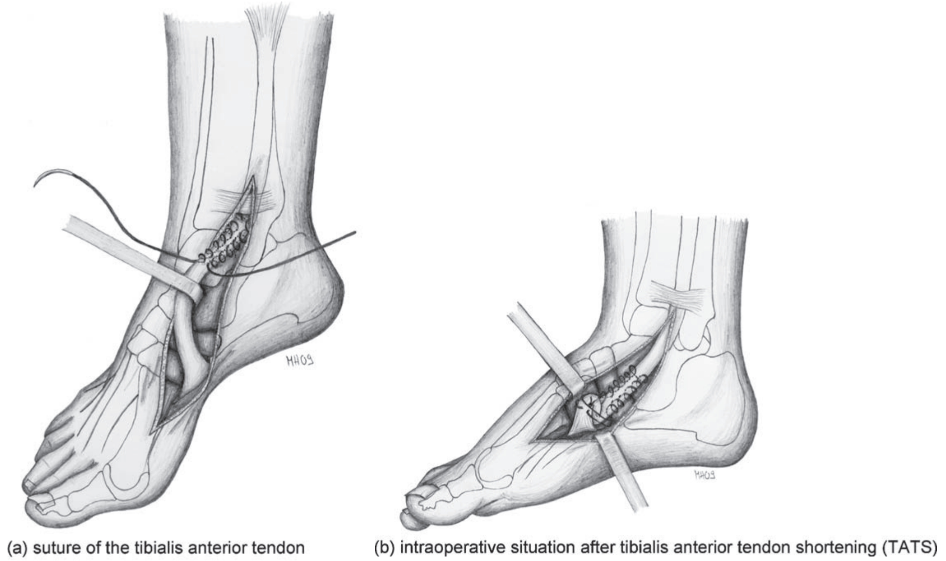


Fig. 1. (a) Suture of the tibialis anterior tendon. (b) Intraoperative situation after tibialis anterior tendon shortening (TATS).

Table 1. Spasticity (S) and manual muscle strength (M) of medial gastrocnemius muscle and tibialis anterior muscle pre- and postoperatively.

	Preoperative	Postoperative	*p*-Value
Group I (hemiplegia, *n* = 21, 21 operated legs)			
S			
Gastrocnemius (range)	2.27 ± 1.39 (0–5)	0.79 ± 0.66 (0–2)	<0.0001#
Tibialis anterior (range)	0.80 ± 0.73 (0–2)	0.29 ± 0.46 (0–1)	0.0099#
M			
Gastrocnemius (range)	3.46 ± 1.06 (2–5)	3.83 ± 1.00 (3–5)	0.2547
Tibialis anterior (range)	3.55 ± 1.06 (1–5)	4.10 ± 0.92 (2–5)	0.0808
Group II (diplegia and quadriplegia, *n* = 8, 9 operated legs)			
S			
Gastrocnemius (range)	1.67 ± 1.36 (0–4)	0.53 ± 0.52 (0–1)	0.0320#
Tibialis anterior (range)	0.69 ± 0.53 (0–1)	0.39 ± 0.47 (0–1)	0.2125
M			
Gastrocnemius (range)	3.47 ± 1.25 (2–5)	4.00 ± 1.19 (3–5)	0.3723
Tibialis anterior (range)	3.67 ± 1.16 (2–5)	4.36 ± 0.93 (2–5)	0.1797

Values are mean ± 1 standard deviation.
Statistically significant.

All patients/parents gave written consent for research purposes in accordance with local ethical committee requirements. The study was performed according to the declaration of Helsinki (World Medical Association).

Statistics

Paired data were assessed using the paired *t*-test or Wilcoxon's matched-pairs signed ranks test, and sequential data were assessed using repeated analysis of variance (ANOVA) with Bonferroni post hoc analysis (SPSS[1] software, Version 15.0; SSPS Inc. Headquarters, Chicago, USA). A power analysis was not carried out.

Results

Spasticity of the gastro-soleus muscles was significantly reduced for patients of both groups postoperatively. For the tibialis anterior muscle spasticity was significantly reduced only for patients of group I, but not for patients of group II (Table 1) postoperatively.

Manually measured strength of the gastro-soleus and tibialis anterior muscles improved postoperatively, compared to pre-operative values. However, this improvement was not statistically significant for patients of either group (Table 1). Nevertheless, active dorsiflexion was present postoperatively in 93% (27 of 29) of the patients.

Table 2 shows the temporal-spatial parameters, the MAP for pelvic tilt, pelvic obliquity, pelvic rotation, hip flexion, hip abduction, hip rotation, knee flexion, ankle dorsiflexion and foot progression, the GPS, the GGI, and the GDI compared pre- to postoperatively for both groups. All patients improved. Group I showed a significant difference for the MAP of hip abduction and for ankle dorsiflexion, for the GPS, the GGI, and the GDI. The mean overall GPS was preoperatively 12.58 ± 3.88 and postoperatively 9.28 ± 2.68. The change in GPS was 3.38. The improvement reported here thus represents a reduction of 45% of the difference between pathological and healthy gait patterns. Three patients in this group had an additional adductor lengthening surgery, three had lengthening of tibialis posterior tendon, two hamstrings lengthening, two a femoral derotation osteotomy and a further two had supramalleolar osteotomy.

In group II there was a significant difference of MAP for ankle dorsiflexion and foot progression, of the GPS, the GGI, and the GDI. The mean overall GPS was preoperatively 14.38 ± 1.38 and postoperatively 9.08 ± 1.38. The change in GPS was 5.38.

Table 2. Temporal-spatial parameters, the Movement Analysis profile (MAP), the Gait Profile Score (GPS), the Gillette Gait Index (GGI), and the Gait Deviation Index (GDI).

	Preoperative	Postoperative	*p*-Value	Direction
Group I (hemiplegia, n = 21, 21 operated legs)				
Walking speed (m/s)	1.16 ± 0.18	1.20 ± 0.21	0.4969	
Cadence (steps/min x 100)	1.16 ± 0.18	1.90 ± 0.19	0.9079	
Stride length (m)	1.22 ± 0.12	1.26 ± 0.15	0.2751	
MAP pelvic tilt	7.50 ± 4.65	6.73 ± 3.26	0.5402	Ant. to post.
MAP pelvic obliquity	3.94 ± 2.19	3.80 ± 2.03	0.8388	Up to down
MAP pelvic rotation	9.16 ± 5.07	7.67 ± 4.04	0.3050	IR to ER
MAP hip flex/ext	10.82 ± 5.18	8.54 ± 4.44	0.1381	Flex. to ext.
MAP hip abd/add	6.70 ± 3.02	4.96 ± 2.21	0.0413#	Add to abd
MAP hip rotation	12.41 ± 5.93	10.95 ± 6.42	0.4550	IR to ER
MAP knee flex/ext	11.69 ± 2.95	10.36 ± 3.17	0.1734	Flex. to ext.
MAP ankle dorsiflex/ext	20.64 ± 11.85	9.62 ± 5.48	0.0004#	Plantar to dorsi
MAP foot progression	13.81 ± 6.41	10.84 ± 5.40	0.1163	
GPS	12.47 ± 3.80	9.18 ± 2.55	0.0022#	
GGI	289.51 ± 239.46	132.96 ± 96.24	0.0085#	
GDI	72.05 ± 9.45	82.61 ± 9.24	0.0008#	
Group II (diplegia and quadriplegia, n = 8, 9 operated legs)				
Walking speed (m/s)	1.12 ± 0.06	1.17 ± 0.06	0.5282	
Cadence (steps/min x 100)	2.16 ± 0.08	2.09 ± 0.08	0.5609	
Stride length (m)	1.06 ± 0.07	1.12 ± 0.07	0.4922	
MAP pelvic tilt	6.16 ± 1.35	7.76 ± 1.35	0.4135	Post. to ant.
MAP pelvic obliquity	3.21 ± 0.36	2.42 ± 0.36	0.1416	Up to down
MAP pelvic rotation	7.98 ± 1.18	6.22 ± 1.18	0.3081	IR to ER
MAP hip flex/ext	13.10 ± 2.78	11.97 ± 2.78	0.7768	Flex. to ext.
MAP hip abd/add	6.19 ± 0.77	5.37 ± 0.77	0.4593	Add to abd
MAP hip rotation	14.96 ± 2.76	10.93 ± 2.76	0.3177	IR to ER
MAP knee flex/ext	18.78 ± 2.82	11.61 ± 2.82	0.0911	Flex. to ext.
MAP ankle dorsiflex/ext	22.83 ± 2.52	7.21 ± 2.52	0.0004#	Plantar to dorsi
MAP foot progression	13.42 ± 1.81	6.97 ± 1.81	0.0224#	
GPS	14.31 ± 1.33	9.00 ± 1.33	0.0120#	
GGI	468.133 ± 97.20	131.02 ± 97.20	0.0260#	
GDI	65.86 ± 3.57	81.31 ± 3.57	0.0075#	

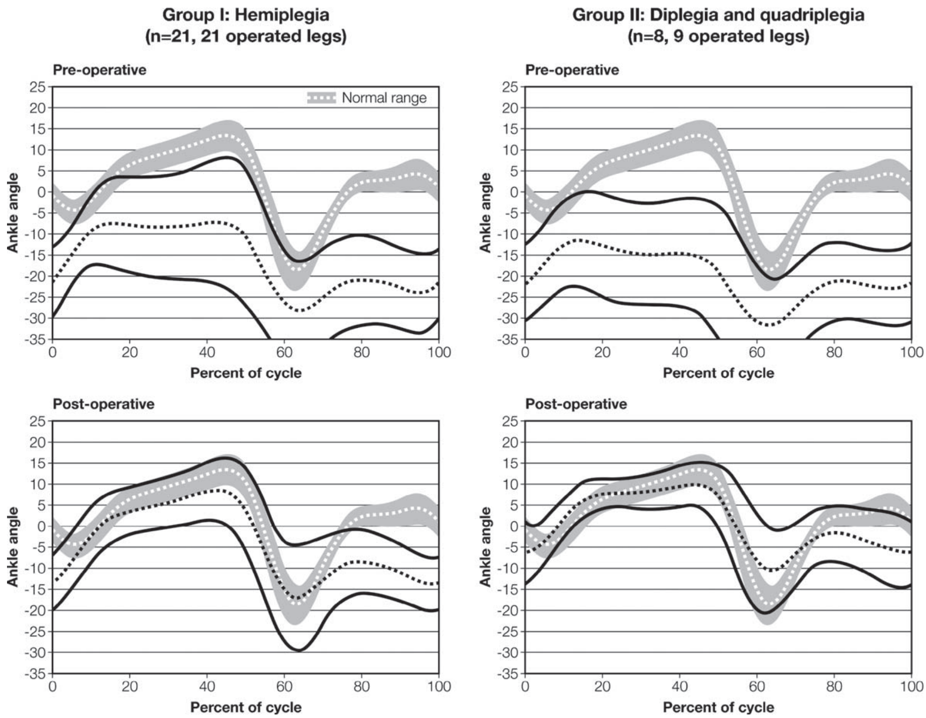


Fig. 2. Ankle kinematics pre- and postoperatively for group I and II.

The improvement reported here thus represents a reduction of 58% of the difference between pathological and healthy gait patterns. One patient in this group had an additional femoral derotation osteotomy, one patient had bilateral and one unilateral hamstring lenthening. The functional correction was good enough to allow walking without orthotics in all patients.

Fig. 2 shows the pre- and postoperative ankle kinematics for all patients.

Table 3 displays the graphs of the preoperative and postoperative MAP's, GPS's, GGI's and GDI's of a 14-year-old patient with right sided hemiplegic CP. He had TAL in combination with TATS and lengthening of the tibialis posterior tendon on the right side. He improved in all scores and his gait pattern almost normalised 12 months after surgery.

Before surgery all patients had a toe walking gait pattern and postoperative 31% of the patients achieved a heel-toe gait or at least a flat-foot contact. The postoperative position of the affected foot at initial contact was for 4 patients (13.8%, 1 patient from group I and 3 patients from group II) a heel-toe-gait, for 5 patients (17.2%, 3 patients from group I and 2 patients from group II) a plantargrade foot, and for 20 patients (69%, 17 patients from group I and 3 patients from group II) a toe-heel-gait.

No surgical complications occurred in the reported series.

Discussion

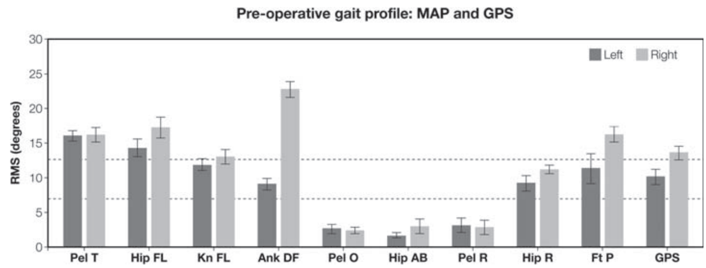
Structural shortening of the gastro-soleus complex is a frequent problem in CP, and several surgical approaches to gain length are described. Jahn et al. [27] compared calf muscle-tendon lengths before and after TAL and gastrocnemius lengthening for equinus in CP and idiopathic toe walking (ITW). Preoperatively muscle-tendon lengths were significantly shorter for the TAL group compared to the Vulpius procedure group. Postoperatively the lengths were not significantly different between the surgeries. There were no significant differences between CP and ITW patients or indications that the surgery affected the groups differently. These studies did not address the tibialis anterior muscle.

Muscle contractures in cerebral palsy are often present for a long time before surgical correction. During this period the agonist is short whereas the antagonist becomes stretched. This stretch contributes to muscle weakness as with overly long tendon-muscle units the muscle becomes ineffective. The muscle contraction unit cannot control the range of motion of the joint anymore. For this reason agonist lengthening (TAL) was combined with antagonist shortening (TATS) at the ankle level. Our study focuses on the functional short-term outcome after this combined procedure for correcting spastic equinus in CP and avoiding a drop foot deformity.

Spasticity of the gastro-soleus muscle was significantly reduced in all patients when compared pre- and post surgery without a change in muscle strength. To set the correct muscle length is challenging, and our surgical method appears to be safe in avoiding over-lengthening and hence weakening of the muscle. Surprisingly, spasticity was also reduced in the tibialis anterior muscle in group I although the muscle was tightened. This effect can either be due to the rebalancing of agonist and antagonist muscle strength or due to the improvement of gastro-soleus spasticity as a trigger.

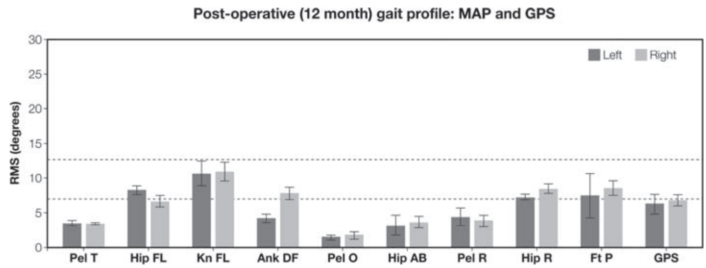
Muscle strength of gastro-soleus and tibialis anterior muscles remained unchanged. Manual muscle test during clinical assessment, however would not necessarily represent muscle function during gait. This clearly improved as 27 out of 29 patients showed active dorsiflexion of the ankle during gait postoperatively. None of the muscles with TATS deteriorated functionally after surgery and one out of three of all patients had a heel-toe gait or at least a flatfoot position at initial contact. This functional result was somewhat disappointing as active dorsiflexion was present in almost all patients. A study of dynamic EMG when walking with and without functional AFOs showed that the activity pattern remained unchanged in spite of gait improvement [28]. It appears that the underlying neurological pattern remains unchanged in spite of the functional corrections.

Table 3. Single case of a 14-year-old patient with right sided hemiplegic CP. TAL and TATS were combined with lengthening of the tibialis posterior tendon.



	Pel T	Pel O	Pel R	Hip Fl	Hip Ab	Hip R	Kn Fl	Ank DF	Ft P	GPS	GGI	GDI
Left	16.1	2.6	3.2	14.3	1.7	9.2	11.9	9.1	11.3	10.1	61.5	76.8
SD	0.8	0.6	1.0	1.3	0.4	1.1	0.9	0.8	2.1	1.1	9.6	1.4
Right	16.2	2.4	2.8	17.2	3.0	11.3	13.0	22.7	16.3	13.6	260.0	67.0
SD	1.1	0.5	1.1	1.5	1.0	0.5	1.0	1.1	1.2	1.0	23.5	1.3

Postoperative MAP, GPS, GGI and GDI



	Pel T	Pel O	Pel R	Hip Fl	Hip Ab	Hip R	Kn Fl	Ank DF	Ft P	GPS	GGI	GDI
Left	3.5	1.4	4.4	8.2	3.2	7.3	10.7	4.2	7.5	6.2	51.0	93.9
SD	0.4	0.4	1.3	0.6	1.5	0.4	1.8	0.6	3.2	1.4	17.6	2.8
Right	3.4	1.8	3.8	6.7	3.7	8.4	11.0	7.8	8.6	6.8	88.0	94.2
SD	0.2	0.5	0.8	0.9	0.8	0.7	1.4	0.9	1.0	0.9	10.9	1.7

TATS is technically easy, quick, and safe (we did not see any complications) to perform and the gait pattern is improved considering the more general outcome parameters. We recommend the combined TATS-TAL surgical procedure followed by intensive physiotherapy to improve foot control. A long-term follow up is necessary to assess long-term correction of gait.

In group I there was a significant difference when comparing the MAP for hip abduction, the MAP for ankle dorsiflexion, the GPS, the GGI, and the GDI pre- and postoperative surgery. The difference in the MAP for hip abduction is probably poorly related to TATS and TAL, as three patients of this group had additional adductor lengthening. Patients of group II, in spite of the small number and different types of CP, improved considering MAP for ankle dorsiflexion and for foot progression, GPS, GGI, and GDI significantly. They seemed to respond even better to the surgical corrections than patients of group I.

Outcome measurement of surgery in CP is difficult. The median value of the GPS for healthy children is 5.28. The improvement reported here thus represents a reduction of 45% for patients of group I, and 58% for patients of group II of the difference between pathological and healthy gait patterns. In our investigation the overall gait pathology was measured by the GPS. To our knowledge this is the first study using MAP and GPS for outcome measurement in children with CP after surgical treatment of spastic equinus.

The limitations of this study are the short-term follow up, lack of a non-operative control group and the retrospective design. However, no data were missing and the data were recorded according to a defined regimen. A long term follow up of this cohort, however, is necessary especially to investigate the recurrence rate of equinus and the function of the tibialis anterior muscle.

We conclude that TATS in combination with TAL in spastic equinus in CP is a safe procedure and improves but not fully corrects foot positioning during gait. Hence, for the treatment of spastic equinus in CP we recommend shortening of the elongated antagonist (TATS) in combination with lengthening of the short agonist (TAL) for achieving optimal postoperative function.

Acknowledgements

The authors wish to thank the Swiss National Science Foundation (SNF) for financially supporting this study, Dr. Monika Horisberger, MD for drawing Fig. 1(a) and (b), and Mary Sheedy for the help in the preparation of this manuscript.

Conflicts of interest statement

There are no conflicts of interest and no benefits in support of the study.

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Chapter 4

Long-term follow up after tibialis anterior tendon shortening in combination with Achilles tendon lengthening in spastic equinus in cerebral palsy

Michèle Kläusler, Bernhard Maria Speth, Reinald Brunner, Oren Tirosh,
Carlo Camathias and Erich Rutz

Abstract

Using Tibialis Anterior Shortening (TATS) in combination with Achilles Tendon Lengthening (TAL) to treat spastic equinus in children with cerebral palsy (CP) was described in 2011. Short-term results have indicated a good outcome, especially an improvement of the drop foot in swing phase and the correction of equinus in stance phase. The aim of this study was to analyse the results of the long-term follow-up and to determine the relapse rate of TATS and TAL.

The kinematics of the sagittal, frontal and transversal planes were measured by using instrumented 3D gait analysis at three defined time points and then described using the Gait Profile Score (GPS) and Movement Analysis Profile (MAP). The data was exported into Gaitabase and then the preoperative (T0), short-term (T1) and long-term (T2) follow-up data was statistically compared.

23 patients (mean age at index-surgery = 14.9 years) were included, there was a mean follow-up time of 5.8 years. 3 children (13%) have shown a relapse. The data of 12 children with spastic hemiplegia (12 legs), as well as 8 children with spastic diplegia (10 legs) has been analysed. There has been a significant ($p < 0.05$) improvement in GPS and MAP for ankle dorsiflexion (describes equinus and drop foot) of the operated legs versus not operated legs.

TATS in combination with TAL shows a satisfactory long-term result after 5.8 years in the correction of fixed equinus and drop foot in children with CP. Postoperatively all subjects were able to walk without an AFO.

Introduction

Among all deformities in children with cerebral palsy (CP) equinus is the most common one [1]. There are conservative and surgical treatment options available. The conservative therapy includes physiotherapy [2], ankle foot orthoses (AFO), castings [3] and injections of botulinum toxin type A [4,5]. Recent studies showed, that the age at time of surgery plays an important role in the development of recurrences [6]. As a consequence, surgery should be evaluated well and, if possible, conservative treatment options should be considered in a young child.

There are several different surgical procedures for the treatment of spastic equinus. The main concept of these methods is to lengthen the gastrocnemicus-soleus muscle-tendon complex. After surgery it is important to continue with the conservative treatment in order to avoid recurrence.

Many surgical procedures have been described over the last few decades. A systematic review by Shore et al. in 2010 [6] has summarized and grouped the outcomes of ten different procedures by anatomic zones. It describes Zone 1 as starting at the gastrocnemius origin and ending at the most distal fibres of the medial belly of the gastrocnemius. Zone 1 procedures include the Baumann [7] and the Strayer [8] distal gastrocnemius recession. Zone 2 is between the distal gastrocnemicus belly and the end of the soleus muscle fibres, procedures belonging to this group are Baker [9] and Vulpius [10] gastrosoleus aponeurotic lengthening. The Achilles tendon is described as Zone 3 including all forms of lengthening of the Achilles tendon, one of them is the Z-tendo-Achilles lengthening (TAL).

In 2011 Rutz et al. published a new concept about the tibialis anterior tendon shortening (TATS) in combination with Achilles tendon lengthening (TAL) in spastic equinus [11]. In manual muscle tests during clinical assessment there was no change in muscle strength of the gastro-soleus and tibialis anterior muscle at short-term. Nevertheless 27 out of 29 patients showed active dorsiflexion of the ankle postoperatively, which was absent in the preoperative test. In 2016 Tsang et al. [12] confirmed favorable outcomes in a similar patient population with a mean follow-up time of 17.9 months.

The aim of our study is to perform a retrospective analysis of all ambulatory children with CP with Gross motor function classification system (GMFCS) level I–II [13] and spastic equinus, equivalent to fixed equinus, to evaluate the long-term clinical and kinematic outcomes using the Movement Analysis Profile (MAP) and the Gait Profile

Score (GPS) [14]. In addition we would like to report the relapse rate of TATS and TAL and the clinical results after TATS in combination with a TAL.

Materials and methods

All ambulatory children (GMFCS level I and II) with unilateral or bilateral spastic CP and two postoperative follow-ups (T1 (1–2 years) and T2 (at least 3 years)) were included.

In clinical examination there was no active ankle dorsiflexion (over 0°) was not possible in any of the patients preoperatively. The exclusion criteria for the study were a diagnosis other than CP, dystonic or mixed movement disorder, Botulinum toxin A injections in the previous 6 months, GMFCS level III or higher.

All patients received a preoperative (T0) three-dimensional gait analysis and a clinical assessment, as well as two postoperative a (T1 (1–2 years), T2 (at least 3 years)) analysis and clinical assessments.

Part of the clinical assessment are the examination of the active ankle dorsiflexion, the passive range of motion (RoM), spasticity according to the modified Ashoworth/ Bohannon scale [15] (scale: 0–4), and the manual muscle strength test ([16,17]) (scale: 0–5) of the dorsi- and plantarflexors. The instrumented gait analysis included kinematics, kinetics and dynamic surface electromyography (EMG). To capture the data a motion capture system (6 camera VICON 460 system), Oxford Metrics Ltd., UK9, 2 force plates (Kistler Instrumente AG, Winterthur, Switzerland) and an 8 channel surface EMG system (Neurodata GmbH, Vienna, Austria) has been used. The Helen Hayes Marker set [16] has been used and six trials were recorded. In addition the anthropometric data has been recorded in order to achieve appropriate scaling.

From the 3D gait data there have been calculations of the temporalspatial parameters (cadence, stride length, and walking speed), the Movement Analysis Profile (MAP) and the Gait Pofile Score (GPS) [14] for all patients pre- and postoperatively for group I (unilateral CP) and II (bilateral) separately, as well as for operated and non-operated legs.

Surgery and postoperative treatment

The surgeries and postoperative treatment were performed as described in the previous study of Rutz et al. [11]. Before performing TAL a test injection with botulinum toxin A in the gastro-soleus muscles was performed to exclude all patients who after the injection showed a deterioration by weakening this muscle [18]. In the remaining patients TAL was performed as described in the previous study by Rutz et al. [11].

Postoperatively a cast was applied in plantigrade (unilateral CP, group I) or mild equinus position (bilateral CP, group II) for 6 weeks postoperatively. If there was no active ankle dorsiflexion after cast removal, the child was provided with a hinged ankle foot orthosis (AFO) with plantarflexion block.

Statistics

Paired t-test or Wilcoxon's matched-pairs signed rank test have been used to assess paired data. Sequential data were assessed using repeated analysis of variance (ANOVA) with Bonferroni post hoc analysis (SPSS® software, Version 42.0; SSPS Inc. Headquaters, Chicago, USA; STATISTICA®, StatSoft Inc.).

Written consent for research purposes in accordance with the local ethical committee requirements has been given by all patients/parents.

The study was performed according to the declaration of Helsinik (World Medical Association).

Results

23 patients with 25 legs, who underwent TATS in combination with TAL between April 2004 and April 2011 and had 2 follow-ups (T1 = 1–2 years, T2 at least 3 years) were included. 3 children with hemiplegia (13%) have shown a relapse (one at T1, two at T2) and therefore they have been excluded from the analysis. The relapses were 2 female (age 19.3 years, 16.2 years) patients and 1 male (age 13.6 years). All of the children with relapses were hemiplegic. Group I consists of 12 children with spastic hemiplegia (12 operated legs, 12 non-operated legs), group II of 8 children with spastic diplegia (10 operated legs, 6 non-operated legs). The mean age at index surgery was 14.9 ± 4.0 years (Group I: 13.3 ± 3.0 years, Group II: 16.6 ± 5.1 years).

Group I included 3 female and 9 male patients. Group II included 4 female and 4 male patients.

The short-term (T1) follow-up time was 1.3 years, and the long-term (T2) follow-up time 5.8 ± 2.1 years (Group I: 5.67 ± 1.7 years, Group II: 5.75 ± 2.8 years).

The mean height at T0 was 153.8 ± 16.5 cm (Group I: 154.8 ± 15.9 cm, Group II: 153.1 ± 18.3 cm) and the mean weight 49.3 ± 16.3 (Group I: 51.2 ± 17.1 kg, Group II 46.5 ± 15.7 kg). At T2 the mean height was 167.5 ± 8.5 cm (Group I: 172.0 ± 6.4 cm, Group II: 161.4 ± 7.2 cm) and the mean weight 63.7 ± 16.8 kg (Group I: 72.9 ± 15.7 and Group II: 51.1 ± 7.8 kg).

Preoperative 8 children had GMFCS level I, 12 children GMFCS level II (GMFCS level in group I: I = 7, II = 5, Group II: I = 1, II = 7). At T2 the GMFCS level in group I was I in 10 and II only in 2 patients, and in group II GMFCS level I was shown in 4 patients and level II in the remaining 4 patients. Overall this means that 6 (30%) patients at T2 were walking at a better level.

14 children had foot corrective surgery only (TATS and TAL) and 5 children (2 children in group I and 3 in group II) had both foot corrective surgery including TATS and TAL and surgeries on a different anatomical level. These surgeries included in group I in one patient tenotomy of semitendinousos muscle and in one child varus derotation osteotomy of the proximal femur. In group II the additional surgeries were lengthening of tibialis posterior in 3 patients and one of them also distal femoral osteotomy.

Preoperatively all children walked with AFO. Postoperatively (at T2) there was no need for AFO's in all patients.

Table 1 shows that spasticity of the gastro-soleus muscle was significantly ($p < 0.05$) reduced for short- and long-term follow-up of group I and significantly only for long-term follow-up of group II. The spasticity of the tibialis anterior muscle was only significantly reduced in the long-term follow-up of group II, but not for any others.

Manual strength of the gastrosoleus and tibialis anterior improved postoperatively at short-term and then deteriorated again long-term. However, there was no statistical significance in both groups.

In all 20 patients clinical assessed active dorsiflexion was possible at T2.

In Table 2 MAP for pelvic tilt, pelvic obliquity, pelvic rotation, hip flexion, hip abduction, hip rotation, knee flexion, ankle dorsiflexion and foot progression as well as the GPS is shown. The two postoperative follow-ups (T1/T2) were statistically compared to the preoperative examination. Looking at both groups together there is a significant improvement ($p < 0.05$) in GPS: 13.3 ± 4.4 preoperative (at T0) compared to 9.7 ± 3.2 (at T1) and 10.4 ± 2.7 (at T2) postoperative. For MAP for ankle dorsiflexion significant difference can be shown: 18.3 ± 10.3 preoperperative (at T0) compared to 9.0 ± 3.4 (at T1) and 9.0 ± 2.7 (at T2). There has been no significant difference for any other parameters. When looking at Group I only, there is only a significant difference of MAP for ankle dorsiflexion, but not for GPS. In comparison in Group II there is a significant improvement of GPS at T1 and T2 as well.

Table 3 There is also a significant improvement ($p < 0.05$) in GPS of the operated legs: 13.3° ± 4.4° (at T0), 9.7° ± 3.2° (at T1) and 10.4° ± 2.7° (at T2) versus not operated legs: 9.7° ± 3.2° (at T0), 8.7° ± 3.3° (at T1) and 8.7° ± 2.5° (at T2). A statistically significant improvement in MAP for ankle dorsiflexion (describes equinus and drop foot) of the operated leg: 18.3° ± 10.3° (at T0), 9.0° ± 3.4° (at T1), and 9.0° ± 2.7° (at T2) versus not operated leg: 7.5° ± 2.2° (at T0), 6.5° ± 1.9° (at T1) and 6.9° ± 2.5° (at T2) was shown.

Fig. 1 shows ankle kinematics pre- and postoperatively for group I and II.

Table 1. Spasticity (S) and manual muscle strength (M) of medial gastrocnemius muscle and tibialis anterior muscle pre- (T0) and postoperatively (T1, T2).

	Preoperative (T0)	Postoperative (T1)	p-Value (compared to T0)	Postoperative (T2)	p-Value (compared to T0)
GroupI(hemiplegia,n=12,12operatedlegs)					
Spasticity					
Gastronemius (range)	1.8 ± 1.3 (0–4)	0.6 ± 0.5 (0–1)	0.013*	0.7 ± 1.1 (0–3)	0.023*
Tibialis anterior (range)	0.4 ± 0.5 (0–1)	0.2 ± 0.0 (0–1)	0.250	0.2 ± 0.0 (0–1)	0.250
Muscle strength					
Gastronemius (range)	3.9 ± 0.9 (2-5)	3.9 ± 0.8 (2-5)	1.000	3.5 ± 1.2 (1-5)	1.000
Tibialis anterior (range)	4.0 ± 0.9 (3-5)	4.5 ± 0.7 (3-5)	0.320	4.2 ± 0.6 (3-5)	0.328
Group II (diplegia, n = 8, 10 operated legs)					
Spasticity					
Gastronemius (range)	1.9 ± 1.1 (0–3)	1.0 ± 0.7 (0–2)	0.087	0.8 ± 0.8 (0–2)	0.026*
Tibialis anterior (range)	0.7 ± 0.5 (0–1)	0.4 ± 0.5 (0–1)	0.063	0.1 ± 0.3 (0–1)	0.029*
Muscle strength					
Gastronemius (range)	3.1 ± 1.1 (2-5)	3.2 ± 1.0 (2-5)	1.000	2.7 ± 1.1 (2-5)	1.000
Tibialis anterior (range)	3.8 ± 1.0 (2-5)	4.2 ± 0.9 (2-5)	0.965	4.0 ± 0.7 (2-5)	1.000

Values are mean l ± standard deviation.
*Statistically significant.

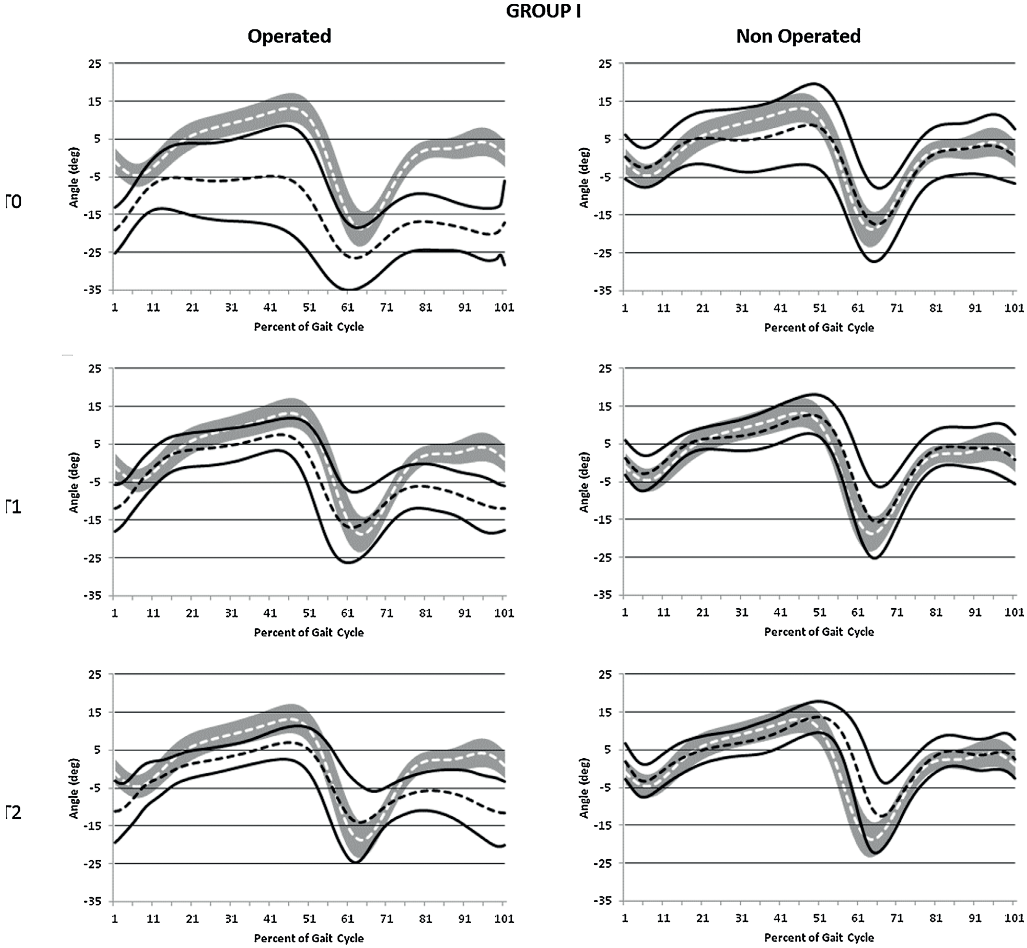

Fig. 1. Ankle kinematics pre- and postoperatively for group I and II pre- (T0) and postoperatively (T1, T2).

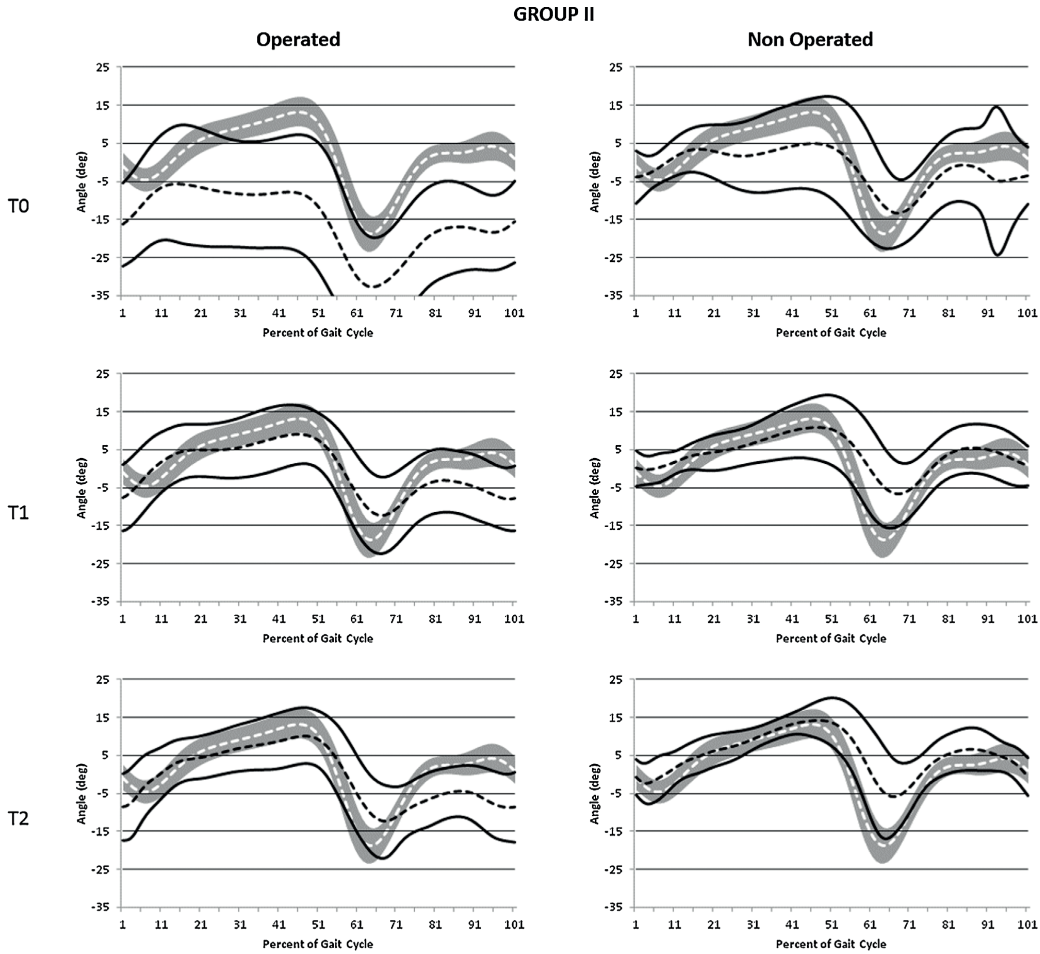

Fig. 1. (*continued*).

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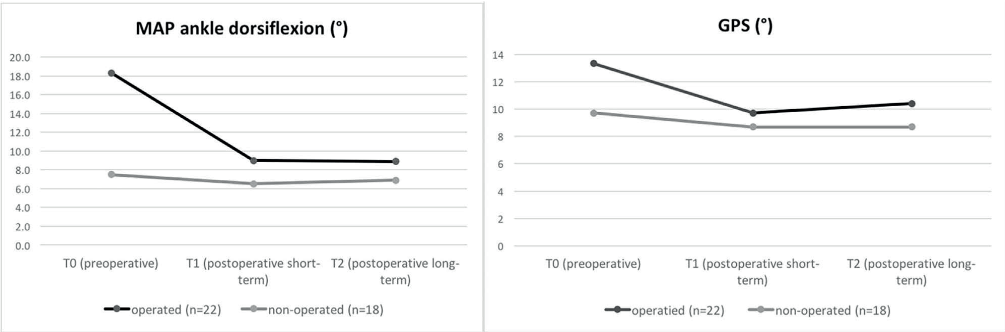
Table 2. The Movement Analysis profile (MAP) and the Gait Profile Score (GPS) for group I and II pre- (T0) and postoperatively (T1, T2).

	Preoperative (T0)	Postoperative (T1)	*p*-Value	Postoperative (T2)	*p*-Value
Group I + II					
MAP pelvic tilt	7.9 ± 3.9	8.9 ± 6.5	1.000	9.3 ± 6.9	1.000
MAP pelvic obliquity	3.6 ± 1.5	3.2 ± 1.5	1.000	3.3 ± 1.4	1.000
MAP pelvic rotation	9.3 ± 4.2	8.0 ± 3.8	0.927	8.4 ± 4.2	1.000
MAP hip flex/ext	10.6 ± 5.6	10.6 ± 6.0	1.000	10.8 ± 5.1	1.000
MAP hip abd/add	5.5 ± 2.5	5.2 ± 2.2	1.000	4.8 ± 1.9	0.846
MAP hip rotation	15.2 ± 10.1	9.8 ± 7.3	0.830	13.04 ± 6.2	1.000
MAP knee flex/ext	15.2 ± 8.2	11.5 17- 5.4	0.233	13.0 ± 6.1	0.822
MAP ankle dorsiflex/ext	18.3 ± 10.3	9.0 ± 3.4	0.00*	8.9 ± 2.7	0.00*
MAP foot progression	14.6 ± 8.6	10.3 ± 4.9	0.910	10.0 ± 5.2	0.590
GPS	13.3 ± 4.4	9.7 ± 3.2	0.003*	10.4 ± 2.7	0.021*
Group I					
MAP pelvic tilt	6.8 ± 3.1	6.4 ± 3.5	1.000	7.4 ± 4.1	1.000
MAP pelvic obliquity	3.3 ± 2.2	3.1 ± 1.6	1.000	3.2 ± 0.9	1.000
MAP pelvic rotation	8.4 ± 5.6	6.7 ± 2.9	0.850	6.6 ± 3.5	0.780
MAP hip flex/ext	9.6 ± 6.2	9.4 ± 5.8	1.000	10.3 ± 4.9	1.000
MAP hip abd/add	5.2 ± 3.0	5.2 ± 2.7	1.000	4.0 ± 1.3	0.687
MAP hip rotation	11.5 ± 6.9	9.1 ± 4.3	0.998	13.0 ± 6.4	1.000
MAP knee flex/ext	12.2 ± 7.4	9.5 ± 4.6	0.772	11.5 ± 4.3	1.000
MAP ankle dorsiflex/ext	17.9 ± 9.5	9.1 ± 2.9	0.003*	8.9 ± 2.3	0.002*
MAP foot progression	9.8 ± 2.8	8.6 ± 3.2	1.000	8.0 ± 4.0	0.607
GPS	11.1 ± 3.7	8.4 ± 1.9	0.053	9.3 ± 1.9	0.312
Group II					
MAP pelvic tilt	9.1 ± 4.3	12.0 ± 7.8	1.000	10.9 ± 8.1	1.000
MAP pelvic obliquity	4.1 ± 2.0	3.4 ± 1.6	1.000	3.6 ± 2.0	1.000
MAP pelvic rotation	10.5 ± 3.7	9.7 ± 4.3	1.000	10.4 ± 4.4	1.000
MAP hip flex/ext	11.8 ± 4.7	12.1 ± 6.2	1.000	11.6 ± 5.8	1.000
MAP hip abd/add	5.9 ± 1.7	5.1 ± 1.5	0.985	5.4 ± 2.0	1.000
MAP hip rotation	19.8 ± 11.8	10.6 ± 10.0	0.141	13.3 ± 6.5	0.499
MAP knee flex/ext	18.9 ± 8.6	14.0 ± 5.5	0.435	13.8 ± 7.5	0.426
MAP ankle dorsiflex/ext	18.8 ± 11.7	9.0 ± 4.0	0.021*	9.1 ± 3.3	0.027*
MAP foot progression	20.5 ± 9.6	12.4 ± 5.9	0.068	12.3 ± 2.0	0.072
GPS	15.9 ± 3.7	11.2 ± 3.3	0.023*	11.4 ± 1.1	0.038*

Values are mean ± 1 standard deviation. Flex: flexion, ext: extension, add: adduction, abd: abduction, dorsiflex: dorsiflexion, dorsiex: dorsiextension. Units for MAP and GPS are degrees.
*Statistically significant.

Table 3. MAP ankle dorsiflexion and GPS in operated (n = 22) vs. non-operated (n = 18) legs.



Discussion

We found a significant improvement of MAP for ankle dorsiflexion and GPS after an average of 5.8 years follow-up time in both groups. Active dorsiflexion was possible in all patients. 30% of the patients showed an improvement of the GCMFS level at T2. Our results show a recurrence rate of 13% (3 out of 25, all hemiplegic patients), but no overcorrection. All 20 included patients were able to walk without AFO (Ankle foot orthosis) at T2. This therefore supports the positive results of the earlier studies by Rutz et al. [11] and Tsang et al. [12].

Compared to our study Tsang et al. [12] describe that the patients need to have active dorsilfexion pre-operatively. In our study none of the patients had active dorsiflexion over 0° pre-operatively. In comparison to this study, for our patients due to fixed equinus the imparison was greater and therefore active dorsiflexion was not possible in clinical examination.

Shore et al. mention in their study, that given all the difficulties of designing randomised surgical trials, especially in a minor group like children, it is important to design good cohort studies. They define this as improved definition of equinus gait and associated functional problems by using instrumented gait analysis, functional testing and quality of life measures. They also emphasise the importance of describing the type of CP more rigorously (movement disorder, topographical distribution and GMFCS level). The surgeries as well as the postoperative management needs to be described accurately, especially in children with spastic diplegia it is essential to fully describe additional procedures in multi-level surgical protocols.

Shore et al. [6] found calcanael deformity and calcaneal gait in children with spastic hemiplegia averaged only 1% (range 0–7%) compared to 15% in spastic diplegia (range 0–41%). In our study we found no calcaneal deformity and no calcaneal gait in any of our patients. Even though highest rates of calcaneus gait seen in longer follow-up [6,19].

Shore et al. [6] describes in their study, that the outcome in terms of recurrent equinus versus calcaneus in children with hemiplegia versus children with diplegia are so different that they need to be looked at in different studies. This study tried to achieve this by splitting patients into 2 groups and the typical difference could be shown.

During the last few years a few studies with mid- and long-term results after surgical procedures for the treatment of spastic equinus have been published, three of them describing Zone I surgeries in diplegic CP patients. Firth et al. [20] published results after a mean of 7 years after Zone I surgeries for children with spastic diplegic CP (GMFCS level II/III), which in the majority of cases were combined with multilevel surgery, orthoses and rehabilitation. None of the patient developed crouch gait and the recurrence at endpoint was 12.5%. They could also show a significant improvement of MAP at both time points. In a study by Svehlik et al. [21], also long-term results (10 year follow-up) after the Baumann procedure in diplegic children were presented.

They had a rate of overcorrection in 9.5% and a recurrence-rate of 23.8%. Dreher et al. [22] published long-term results after gastrocnemiussoleus intramuscular aponeurotic recession also in spastic diplegic cerebral palsy with a mean follow-up of 9 years. They could show a significant improvement of kinematic gait analysis as well as passive dorsiflexion in clinical examination, significant loss of passive dorsiflexion at time of long-term follow-up, but improvement in gait analysis parameters were maintained. There was a recurrence of 24% of legs and late onset calcaneal gait seen in long-term follow-up in eight legs (10%), four of the eight had a crouch gait.

Several studies have shown that more children with hemiplegia show recurrence [6]. The results of this study show similar results with recurrence in three patients with hemiplegia.

Svehlik et al. [21] mention in their study, that in long-term follow-up there is also an effect of growth and increasing weight. Equinus decreases and ankle dorsiflexion increases as children gain weight and go through the adolescent growth spurt. Not all changes seem to be attributed to surgery. Also in our study there has been an increase in weight and height over time.

Reimers et al. [23] published a study which showed that antagonist function improves when spasticity in the agonist is reduced by tendon lengthening. Average age was 5 years, average follow-up time 14 months.

Davids et al. [24] investigated active ankle dorsiflexor function after plantar flexor surgery in children with cerebral palsy. Active ankle dorsiflexor function in swing phase present in 79% of the extremities prior to ankle plantar flexor surgery, after surgery in 96%. They found no significant change between preoperative and postoperative active ankle dorsiflexor function in swing phase.

In our study clinically there was no active dorsiflexion possible in any of the patients preoperatively, in all patients of group I and II active dorsiflexion was possible postoperatively. Also we found a significant change in MAP for dorsiflexion at T2 in both patient groups. All patients that received TATS in combination with TAL were severe cases of fixed equinus, without active dorsiflexion over 0°. In our opinion especially these patients benefit from TATS in combination with TAL.

Our study has numerous limitations. First, we had a small sample size of only 23 children. Secondly, we lacked control groups of untreated patients or children operated with TAL only. Thirdly, we had to exclude three children from the analysis because of relapse. Fourth, our findings are likely to be relevant only for a specific group of children with CP. Finally there was no measure of function and no quality of live assessment scores included.

Given all of this, besides the need for further research, our study shows satisfactory long-term results after 5.8 years after performing TATS in combination with TAL. To correct spastic equinus deformity in unilateral or bilateral cerebral palsy we recommend TAL in combination with TATS.

Conflicts of interest

There are no conflicts of interest and no benefits in support of this study.

Acknowledgements

We would like to thank Mary Sheedy and Grenville Deasy for the help with the preparation of this manuscript.

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4

Chapter 5

Mulitlevel surgery improves gait in spastic hemiplegia but does not resolve hip dysplasia

Erich Rutz, Elyse Passmore, Richard Baker and H. Kerr Graham

Abstract

Background Multilevel orthopaedic surgery may improve gait in Type IV hemiplegia, but it is not known if proximal femoral osteotomy combined with adductor release as part of multilevel surgery in patients with hip dysplasia improves hip development.

Questions/purposes We asked whether varus derotational osteotomy of the proximal femur, combined with adductor release, influenced hip development in patients with Type IV hemiplegia having multilevel surgery.

Patients and Methods We retrospectively reviewed 11 children and adolescents with Type IV hemiplegia who had a proximal femoral osteotomy due to unilateral hip displacement to correct gait dysfunction between 1999 and 2006. The mean age at the time of surgery was 11.1 years (range, 7 to 16 years). We obtained the Movement Analysis Profile and Gait Profile Score before and after surgery. We also measured the Migration Percentage of Reimers and applied the Melbourne Cerebral Palsy Hip Classification System (MCPHCS). The minimum followup was 2 years 3 months (mean, 6 years 6 months; range, 2 years 3 months to 10 years 8 months).

Results The majority of gait parameters improved but hip development was not normalized. According to the MCPHCS at last followup, no hips were classified as Grade I, two hips were classified as Grade II, and the remainder were Grade III and IV.

Conclusions Unilateral surgery including a proximal femoral osteotomy improved gait and walking ability in individuals with spastic hemiplegic cerebral palsy. However, hip dysplasia persists.

Level of Evidence Level IV, therapeutic study. See Guidelines for Authors for a complete description of levels of evidence.

Introduction

Spastic hemiplegia (SH) is the most common type of cerebral palsy (CP) in large population-based studies [20]. Subjects with spastic hemiplegic CP have a high level of gross motor function, Level I or II according to the Gross Motor Function Classification System (GMFCS) [13, 20, 27]; that is, they are able to walk in the community without assistive devices, which reportedly protects against hip dislocation [19].

Gait patterns in subjects with SH have been classified into four groups by Winters, Gage, and Hicks (WGH) [39]. In Type IV hemiplegia, there is involvement at all three levels: the ankle, knee, and hip. At the hip level, there is increased flexion, adduction, and internal rotation [11, 33]. Clinically there is evidence of increased femoral neck anteversion and there may be associated hip dysplasia [32]. Although not all subjects with hemiplegia can be readily categorized using the WGH system and transitional types may occur, the reliability of the classification is good [9, 30]. Importantly, many clinicians believe the WGH classification provides a useful template on which to base treatment decisions [33].

In a population-based study of children with CP from our state, the prevalence of hip displacement defined as a migration percentage (MP) greater than 30% was 35% [35]. This was similar to the prevalence reported in other population-based studies based on CP registers [5, 17]. Two subjects with SH from this population-based cohort study had hip dysplasia and underwent reconstructive surgery for a prevalence rate of 2% of all individuals with SH at skeletal maturity. However, given that only 19 subjects had Type IV hemiplegia, the prevalence of hip dysplasia in Type IV hemiplegia is much higher (two of 19 [10.5%]) [10].

The treatment of hip displacement in SH is important because such individuals have a normal life expectancy and high demands in comparison to individuals with bilateral CP subtypes. Treatment options for the younger child with SH include injections of botulinum toxin A for spastic equinus and the provision of a suitable ankle-foot orthosis [1, 15]. The older child or adolescent with Type IV SH may benefit from unilateral multilevel surgery based on instrumented gait analysis [25]. We previously found unilateral multilevel surgery, including an external rotation osteotomy of the proximal femur, corrects both sagittal and transverse plane gait deviations, including pelvic retraction [8]. Whether these operations achieve the additional goal of normalizing gait or hip development is not clear. There are currently no studies that address this question in the literature. Hip displacement in hemiplegia is rare and may not be recognized until it becomes symptomatic in adult life [3, 16, 17, 19, 21, 35].

The questions addressed in this study were twofold: (1) does unilateral multilevel surgery improve gait? (2) Does unilateral multilevel surgery normalize hip development?

Patients and Methods

We retrospectively reviewed the records of all 16 children with Type IV hemiplegia who had unilateral multilevel surgery, including an external rotation osteotomy of the proximal femur between January 1999 and December 2006. Eight of the 11 patients have previously been reported in a different format not including Gait Profile Score (GPS), Movement Analysis Profile (MAP), or radiology [8]. The indications for surgery were: (1) deterioration in gait; (2) evidence of contractures and bony torsion on physical examination; and (3) radiographic evidence of hip dysplasia, MP [30%. The contraindications for surgery were: (1) stable gait; (2) no fixed deformities; and (3) normal hip development. During this time, we performed this procedure on a total of 16 patients. For this study, we included patients with: (1) Type IV hemiplegia confirmed clinically and by instrumented gait analysis; (2) hip displacement with MP greater than 30%;

unilateral surgery, which included a proximal femoral osteotomy (PFO) explicitly intended to correct both hip dysplasia and gait dysfunction using tailored combinations of external rotation, varus, and extension; and (4) at least one postoperative gait study and clinical and radiographic followup of more than 2 years. Five children had the surgery but were excluded from the study because they had not had a preoperative gait analysis, leaving 11 children for review. There were six boys and five girls with a mean age of 11.1 ± 2.7 years (range, 7 to 16 years) at the time of surgery. Ten children were GMFCS Level II and one child was GMFCS Level III preoperatively. Four had previous surgical procedures mainly for equinus gait, and one had a PFO at other institutions (Table 1). The minimum followup was 2 years 3 months (mean, 6 years 6 months; range, 2 years 3 months to 10 years 8 months). No patients were lost to followup. No patients were recalled specifically for this study; all data were obtained from medical records and radiographs. This study was approved under the audit provisions of the institution's Ethics in Human Research Committee, Number CA 29012. The study was performed according to the Declaration of Helsinki (World Medical Association).

Each patient had a comprehensive history, a standardized clinical examination, radiography of the pelvis [40], and examination under general anesthesia immediately before surgery to confirm the presence of fixed contractures [6].

All children had an instrumented three-dimensional gait analysis (3D gait analysis) preoperatively, at short-term (1.0 year postoperatively), and at midterm (6.4 years postoperatively) followup. The gait analysis was carried out using a 50-Hz six-camera Vicon 370 system (Oxford Metrics, Oxford, UK) and two force plates (Advanced Mechanical Technology Inc, Newton, MA). Reflective markers were applied to the bony landmarks of the lower limb using a standardized protocol. Euler angles and inverse dynamics were used to calculate kinematics and kinetics using the Vicon Clinical Manager (Oxford Metrics). Patients were asked to walk barefoot in their usual manner along a 10-m walkway. An experienced physical therapist (JR, PT) and biomechanical engineer (RB) were responsible for all data collection using standardized protocols and data entry.

Table 1. Patient demographics, previous surgery, and VDRO or multilevel surgery.

Case number	GMFCS	Side	Age at surgery (years)	Previous surgery	Multilevel surgery
1	II	L	13		VDRO, P Add, Botox to Gastrocsoleus
2	II	L	8		VDREO, P Add, Strayer
3	II	R	11	TAL	VDREO, P Add, RFT t oST, MHS, Strayer
4	II	L	9		VDREO, O Add, Botox to Gastrocsoleus
5	II	R	7	Strayer	VDRO, P Add
6	II	R	15		VDREO, O Add, RFT t oST, M&LHS
7	II	R	13		VDREO, P Add, SMO
8	II	R	10	SMO, POTB	VDREO, P Add, PATB, RFT t oST, MHS, Add, Strayer
9	II	L	10	O Add, RFT	VDREO, P Add, Botox to Gastrocsoleus
10	II	R	10		VDRO, O Add, Strayer
11	III	L	16	VDRO, TAL	VDREO, San Diego, O Add, SMO, TAL

Note: All subjects had a unilateral adductor release combined with ipsilateral VDRO or multilevel surgery; Case Numbers 1, 2, 3, 5, 7, 8, and 9 = percutaneous adductor release (P Add); Case numbers 4, 6, 10, and 11 = open adductor release (O Add); VDRO = varus derotation osteotomy; VDREO = varus derotation extension osteotomy; TAL = tendo-Achilles lengthening; SMO = supramalleolar osteotomy; POTB = psoas over the brim; RFT = rectus femoris transfer; MHS = medial hamstring lengthening; M&LHS = medial and lateral hamstring lengthening.

The MAP and GPS have been developed to summarize the complex information arising from 3D gait analysis. The MAP quantifies the magnitude of an individual subject's gait deviations, across nine individual kinematic variables, over the whole gait cycle [2]. The GPS is a summary statistic of gait, which quantifies the overall deviation of kinematic data relative to normative data [2]. All data were uploaded into GaitaBase, a web-based repository for gait data [36]. The MAP and GPS were calculated for all subjects. The median value of the GPS for healthy children is 5.2° and one SD is 1.3°. All children had a preoperative and short-term (1.0-year postoperative) 3D gait analysis.

Standardized AP pelvic radiographs were analyzed by measuring the MP of Reimers [29], center-edge angle (CEA) of Wiberg [38], the acetabular angle of Sharp [34], and pelvic obliquity [37]. Hip morphology was classified using the Melbourne Cerebral Palsy Hip Classification System (MCPHCS) preoperatively, at baseline, 1 year after surgery, and at most recent followup in all subjects. The MCPHCS is a categorical classification of hip morphology based on both qualitative features as well as quantitative measures (MP) and has reasonable reliability [24].

The procedures selected for improving gait were based on clinical examination, 3D gait analysis, and examination under anesthesia following published guidelines [1, 12, 25, 26]. Procedures included lengthening of the gastrocsoleus for equinus gait (Botox1 injections, gastrocnemius recession, or TAL based on Silfverskiö ld test), hamstring lengthening with transfer for the rectus femoris to semi-tendinosus for flexed-stiff knee gait, and lengthening of the iliopsoas and hip adductors. Percutaneous adductor releases were performed and did not routinely include the iliopsoas lengthening. When indicated, iliopsoas lengthening was performed over the brim of the pelvis. Supramalleolar osteotomy of the tibia was performed in two children with malignant malalignment, ie, an external rotation osteotomy of the proximal femur was combined with an internal rotation osteotomy of the distal tibia. The indications for specific procedures were based on data gathered from kinematics, physical examination in the gait laboratory, and examination under anesthesia immediately before surgery [1, 6, 8].

An intertrochanteric osteotomy of the proximal femur was performed in all subjects and fixed with a 90° or 100° fixed-angle blade plate (Synthes1, Solothurn, Switzerland) [12, 18, 26]. The aim of surgery was to correct the mildly valgus neck shaft angle to a normal range between 120° and 130° by combining 5° to 15° of varus with 25° to 40° of derotation. All femoral osteotomies included external rotation based on clinical and CT measurement of ante-version, transverse plane hip kinematics, and evaluation of the AP hip radiograph [32]. Excessive varus was avoided to minimize abductor weakness and exacerbation of limb length discrepancy. When hip flexion contracture persisted at examination under anesthesia, despite previous or concurrent psoas lengthening, a small amount of extension (10°–20°) was built into the osteotomy by altering the inclination of the seating chisel.

Below-knee plaster casts were used for those subjects who required equinus surgery. Stable fixation was achieved and full weightbearing within 2 weeks of surgery was encouraged in all patients. Casts were removed 6 weeks after surgery and placed by custom-molded, articulated, ankle-foot orthoses. Assistive devices were provided until independent walking was regained. Postoperative physical therapy was provided at a frequency of two to three times per week for 3 to 6 months according to progress [1]. Postoperative progress was monitored by return visits to the gait laboratory for a physical examination and two-dimensional video recording of gait at 3, 6, and 9 months after surgery. At 12 months after surgery, a repeat 3D gait analysis was performed.

All data were normally distributed. We compared pre- and postoperative gait parameters using a paired t-test. We used SPSS1 software (Version 15.0; SSPS Inc, Chicago, IL, USA). A power analysis was not carried out.

Results

We observed improvement of overall gait function. The GPS for the affected legs was 16° ± 2.8° and improved (p = 0.0002) by 6° to 11° ± 3° at the short-term followup 3D gait analysis. Kinematic parameters improved in seven of the nine MAP kinematic domains and the improvements were in the transverse plane for pelvic rotation, hip rotation, and foot progression (Fig. 1; Table 2).

Table 2. A comparison of pre- and postoperative kinematics, affected side only, using the Movement Analysis Profile (MAP) and Gait Profile Score (GPS) (N = 11).

Kinematics	Preoperative	Short-term	p value
MAP pelvic tilt	9.6 ± 3.8	8.8 ± 6.9	0.833
MAP hip flexion	10.1 ± 3.3	11.0 ± 7.6	0.700
MAP knee flexion	16.1 ± 5.9	12.6 ± 4.2	0.124
MAP ankle dorsiflexion	14.7 ± 8.9	10.3 ± 3.8	0.147
MAP pelvic obliquity	3.4 ± 2.0	4.5 ± 2.1	0.239
MAP hip abduction	6.7 ± 2.3	5.7 ± 2.4	0.345
MAP pelvic rotation	13.1 ± 4.9	8.3 ± 3.6	0.016
MAP hip rotation	20.9 ± 10.0	9.3 ± 4.6	0.002
MAP foot progression	26.4 ± 10.9	13.0 ± 8.1	0.003
GPS total	16.0 ± 2.8	10.5 ± 2.9	0.026

Units for MAP and GPS are degrees.

Although a small improvement in MP was found at both short-term and medium-term followup, we observed no changes in the CEA of Wiberg or Sharp's angle (Table 3). Pelvic obliquity averaged 4° ± 5° at baseline and tended to improve to 2° ±

3° at short-term followup with a slight relapse to 2° ± 4° at last followup. According to the MCPHCS (see Appendix 1), all hips on the uninvolved side and none of the hips on the involved side were Grade I at most recent followup. Only two hips on the hemiplegic side were classified as Grade II postoperatively and the rest were Grade III or IV. One hip that was managed by varus derotation osteotomy (VDRO) and a San Diego-type acetabuloplasty improved from Grade IV to Grade II (Fig. 2A–C) [23]. One patient with preoperative Grade II remained postoperatively in the same level (Table 3).

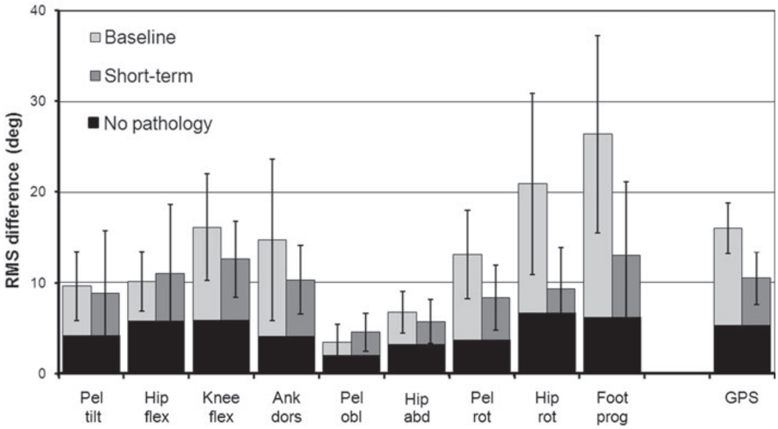


Fig. 1. Movement Analysis Profile (MAP) and Gait Profile Score (GPS) at baseline and short-term followup for the whole cohort. Pel = pelvis; flex = flexion; ank = ankle; dors = dorsiflexion; obl = obliquity; abd = abduction; rot = rotation; prog = progression.

Table 3. Migration percentages for all hips*.

Hip classification				Migration percentage			Followup (years)
Baseline	Short	Medium		Baseline	Short	Medium	
II	II	II		14	12	12	6
III	III	III		16	18	19	10
III	III	III		19	20	20	10
III	III	III		21	18	20	2
III	III	III		22	16	17	6
III	III	III		26	20	18	3
III	III	III		26	27	25	4
IV	III	III		32	25	25	10
IV	III	IV		36	24	31	2
IV	III	IV		39	24	33	9
IV	II	II		57	12	12	8
			Average	25	20	22	6
			SD	8	5	6	3
p (paired t-test from baseline)					0.027	0.025	

* Statistical analysis excludes the one child who had acetabuloplasty as well as varus derotation osteotomy.

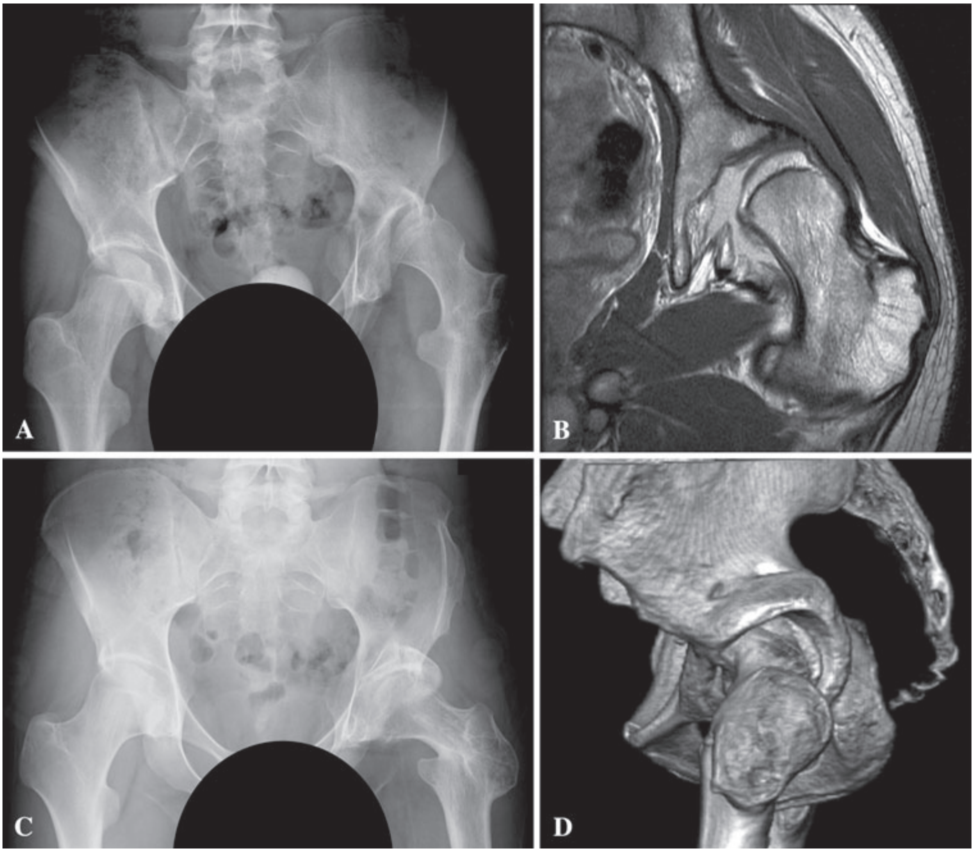


Fig. 2A–D. (A) These are the radiographs of a 16-year-old boy with a left Type IV hemiplegia and painful subluxation of his left hip, advanced femoral head deformity, and severe acetabular dysplasia. At presentation, his mobility had declined from Gross Motor Function Classification System (GMFCS) I to GMFCS III and the left hip was Grade IV according to the Melbourne Cerebral Palsy Hip Classification System (MCPHCS). (B) MRI of the left hip showing cartilage loss and femoral head deformity. (C) Eight-year followup after open reduction/ adductor lengthening, proximal femoral osteotomy (varus, external rotation, and extension), and San Diego Type Pelvic Osteotomy. Pain was completely abolished and he regained independent walking, GMFCS Level II. (D) This three-dimensional CT reconstruction lateral view shows the extension component of the femoral osteotomy and good femoral head containment.

At most recent followup, none of the patients had hip pain on the hemiplegic side. Apart from implant removal, no additional surgeries were performed (Fig. 3A–B). There were no nonunions, delayed unions, or deep infections after VDRO. No patient required a blood transfusion. There were three superficial wound infections, one in an anterior knee incision for rectus femoris transfer and two in the popliteal fossa after hamstring lengthening. These resolved after administration of oral antibiotics.

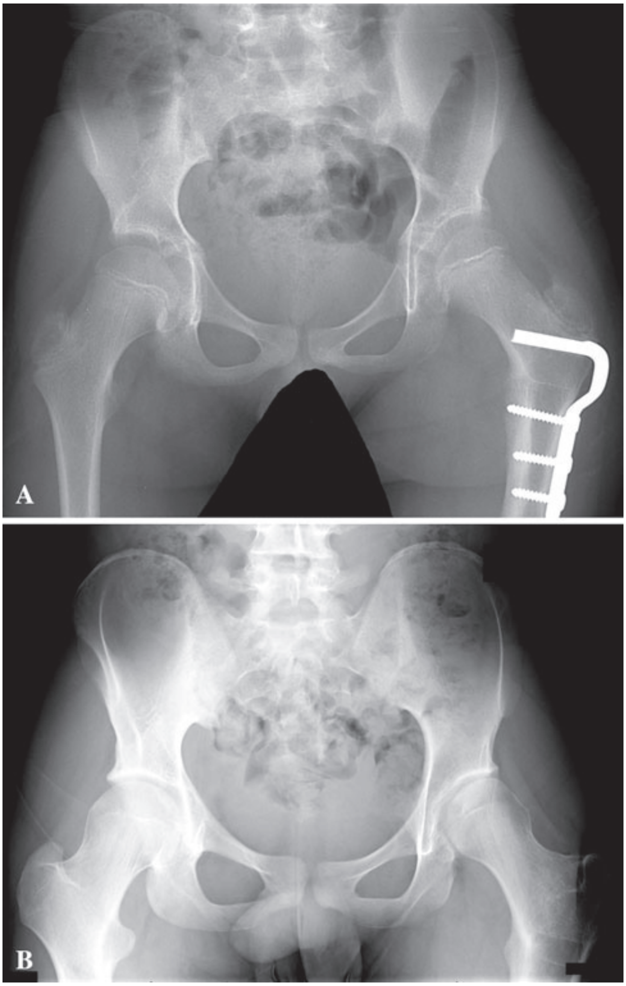


Fig. 3A–B. (A) These are the radiographs of a 9-year-old boy with Type IV left hemiplegia 1 year after left proximal femoral osteotomy, adductor and psoas lengthening, and surgery for equinus deformity. The osteotomy has united and the femoral head is well contained with good acetabular coverage. (B) At skeletal maturity (after removal of the blade plate), Shenton's arch is intact but the femoral head is deformed with a flattened area laterally and there is residual acetabular dysplasia. The left hip migration percentage was 42% and was Grade IV according to the Melbourne Cerebral Palsy Hip Classification System (MCPHCS).

Discussion

The literature suggests both single-level and multilevel surgery improve gait and function in spastic hemiplegia. Single-level surgery, for the management of equinus gait, reportedly improves gait in Type II hemiplegia [14]. In Type IV hemiplegia, several recent reports, using gait analysis, have reported improvement in sagittal and trans-verse plane gait parameters after unilateral, multilevel surgery [4, 8]. We asked whether: (1) unilateral multilevel surgery improves gait and (2) unilateral multilevel surgery normalizes hip development.

We caution readers of the limitations to our study. First, we had a small number of patients. This is a very small subset of patients that has not been previously reported. Our cohort was too small to permit analysis by age at surgery. Surgery at a younger age might run the risk of recurrent limb deformities and regression of the gait improvement component of the surgery. However, surgery at a younger age might encourage more normal acetabular development and provide additional time for monitoring of residual acetabular dysplasia with increased opportunities for pelvic osteotomy before skeletal maturity. However, all included patients were followed with a consistent protocol. Second, we lacked a control group, but MAP and GPS quantify the deviation of kinematic data relative to normative data.

We demonstrated overall improvement in gait using a summary statistic of gait, the GPS. In addition, the use of the MAP shows which kinematic parameters have improved and by how much. From this, some inferences can be made as to which procedures may have been most effective. The focus of our surgery was primarily the correction of gait dysfunction. At the time of presentation, walking, not asymptomatic hip dysplasia, was the patient's and parents' concern. However, we noted mild hip dysplasia in all of our patients and modified the gait correction surgery in an attempt to simultaneously improve gait and correct hip displacement. Although we observed an improvement in the MP, we found no changes in the indices of acetabular dysplasia, the CEA of Wiberg, or acetabular angle of Sharp. MP changes are only relevant for the individual and the mean values for a cohort could only indicate a trend. In addition, none of the hips on the affected side were rated as normal on the Melbourne Cerebral Palsy Hip Scale. The hip that improved most was the only one to have a concomitant pelvic osteotomy. Factors that may have contributed to persisting dysplasia include muscle imbalance, pelvic obliquity, and older age at index surgery. Paradoxically, the pelvic obliquity was always high on the affected side despite some degree of shortening of the affected limb. We believe the pelvic obliquity may be centrally or neurologically mediated and this may explain the lack of response to adductor lengthening. We are unsure of the importance of the residual dysplasia in this population. Patients with hemiplegia have a normal life expectancy and relatively high levels of physical functioning. Longer-term followup will be required to determine if their hip dysplasia will become symptomatic.

Hip dysplasia is common in CP and the prevalence is related to walking ability as determined by the GMFCS but not to movement disorder type [5, 17, 35]. The effects of hip displacement are also related to GMFCS level. The high-risk groups are nonambulatory children with spastic quad-riplegia, GMFCS IV and V [5, 17, 35]. In these patients, pain and fixed deformities that interfere with sitting are the most important issues. Although hip displacement is relatively rare in SH, the effects may be devastating with early onset of pain, gait difficulties, and loss of walking ability

(Fig. 2A–D). As a result of most patients with hemiplegia being at low risk for hip dysplasia, routine screening radiographs are not typically performed. Therefore, the majority of patients did not have initial radiographs until referred for gait analysis. The pathogenesis of hip displacement is different in SH compared with other CP subtypes. Hip displacement develops quite late and may progress slowly and silently until the pubertal growth spurt. Presentation during the pubertal growth spurt may include sudden onset of pain, deterioration in gait, and the need for an assistive device. Increased pelvic obliquity with increased apparent leg length discrepancy may also be features (Fig. 2A–D). We noted a consistent pattern with small leg length discrepancy (LLD) smaller than the apparent LLD and mild pelvic obliquity of 3° to 14° on kinematics (affected side being up and the unaffected side being down). For these reasons, systematic hip surveillance with clinical and radiographic examination at regular intervals is supported in the recent literature [7, 16]. The authors would recommend an initial AP radiograph of the pelvis after the initial diagnosis of Type 4 hemiplegia is made with repeated radiographs every 12 to 24 months until skeletal maturity or until consecutive radiographs have demonstrated normal hip development over a minimum of 2 years.

Although our patients are currently asymptomatic, we radiographically documented substantial acetabular dysplasia in an otherwise high-functioning group of young adolescents. We believe more aggressive management of acetabular dysplasia should be considered in patients with Type IV hemiplegia and hip displacement. Unilateral VDRO, even when combined with adductor and psoas release, improves the migration percentage in these patients but does not result in predictable or complete resolution of acetabular dysplasia. In retrospect, our criteria for acetabular reconstruction, based on MP > 40%, were too lax and this threshold is too high for this CP subtype. However, we believe this threshold is too high, especially in older children, and it may be more appropriate to consider a much lower threshold for pelvic osteotomy, an MP of 20%. Although MP is the most widely used radiographic in CP hip displacement in general, other factors including the acetabular index and the development of the acetabular sourcil should be considered.

Although there is extensive literature on the management of hip displacement in bilateral CP, spastic diplegia, and spastic quadriplegia, there is very little information on the management of hemiplegic hip disease [19, 21, 22, 28]. We presumed correction of hip adduction and flexion contractures combined with a VDRO and appropriate muscle tendon surgery at the knee and ankle would be effective in correcting both the hip displacement and the gait dysfunction in Type IV SH. Fluoroscopic screening before VDRO showed improved femoral head cover and a reduction in MP in all hips. However, although the surgery improved gait, the effects on subsequent hip development were disappointing with a high incidence of residual dysplasia. The VDRO only slightly improved the radiographic parameters (CEA and MP) and did not improve the acetabular angle. The combination of mild pelvic obliquity, which was always high on the affected side, combined with persisting muscle imbalance may have contributed to the persisting hip dysplasia. The only hip that improved by two grades had a concomitant pelvic osteotomy (Fig. 2A–D). Several authors suggest more aggressive management of acetabular dysplasia should be recommended in hemiplegic hip disease [22, 23]. Even mild pelvic obliquity appears to contribute to failure of acetabular development at skeletal maturity (Fig. 3A–B) [3].

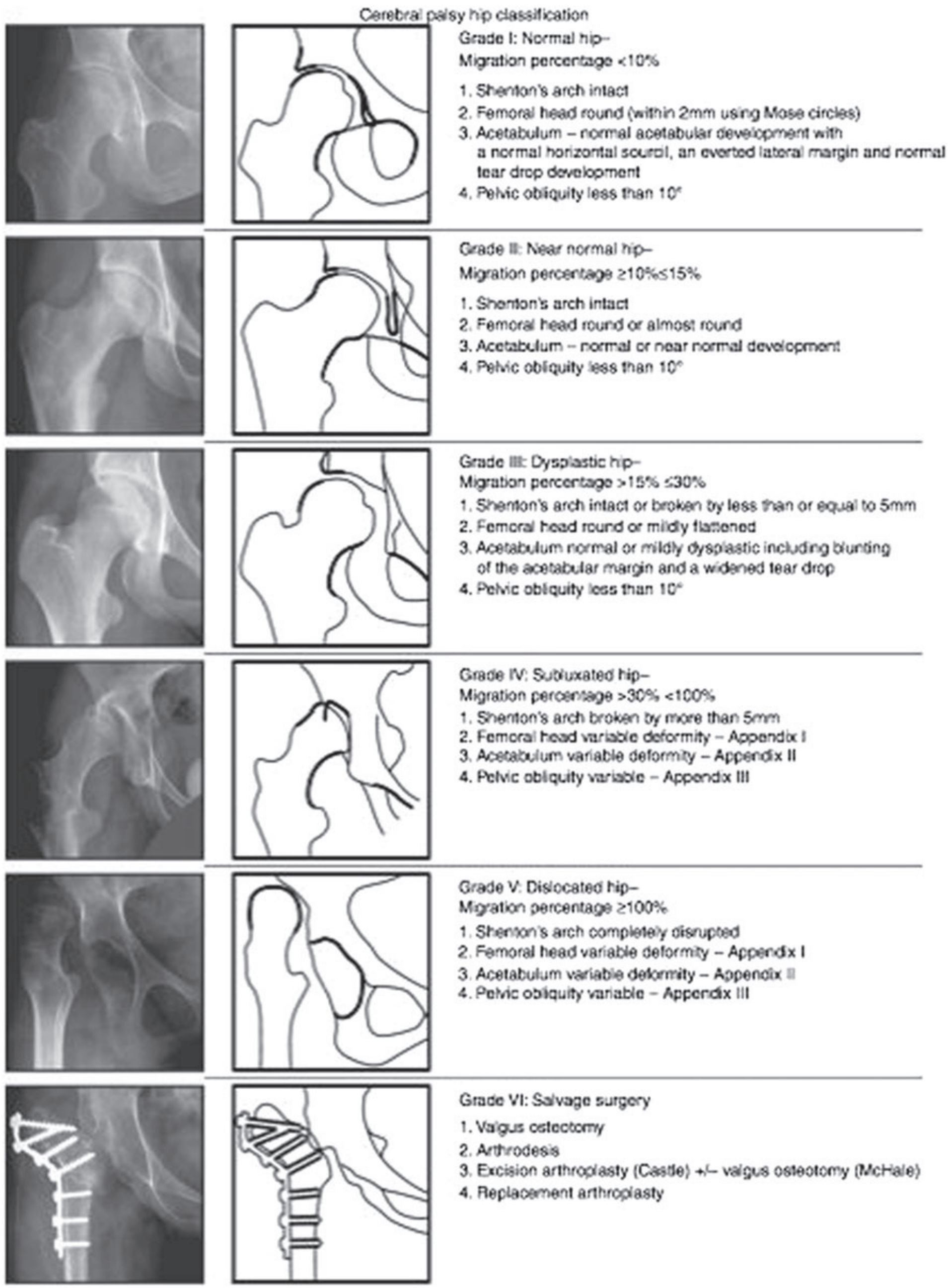
The MCPHCS was introduced to describe hip disease at skeletal maturity across the entire spectrum of CP [24, 31]. We found it a useful tool to discriminate and describe normal hip development on the uninvolved side in individuals with SH as well as a broad spectrum of dysplasia on the affected side [24, 31]. Likewise, the GPS was responsive for detecting clinically meaningful improvements in gait and, in conjunction with the MAP, the kinematic domains where we observed most improvements [2]. The improvements in the sagittal plane domains of the MAP were smaller and insignificant because the ankle and knee gait dysfunction was relatively small because of previous surgery, Botox1, and bracing. We did observe improvements in the transverse plane, secondary to the beneficial effects of VDRO on gait, in combination with additional muscle tendon lengthenings and tendon transfers, when indicated [4, 8].

In conclusion, unilateral orthopaedic surgery, which includes a PFO, is effective in improving gait and walking ability in individuals with spastic hemiplegic CP. However, hip dysplasia persists and although none of our patients were currently symptomatic, we are concerned about the longevity of these hips given normal life expectancy and high levels of function.

Acknowledgments

We thank Mary Sheedy for the help in the preparing and editing the manuscript and the staff of the Hugh Williamson Gait Laboratory for the kinematic data, especially Jill Rodda [JR] and Pam Thomason [PT].

Appendix 1



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Chapter 6

Hip flexion deformity improves without psoas-lengthening after surgical correction of fixed knee flexion deformity in spastic diplegia

Erich Rutz, Mark S. Gaston, Oren Tirosh and Reinald Brunner

Abstract

Background: It is unclear if psoas lengthening surgery is required in the treatment of patients with cerebral palsy (CP) with hip flexion deformity and previous studies show equivocal results with regard to functional outcome.

Methods: This study retrospectively assessed 12 patients with a diagnosis of spastic diplegia who underwent single event multilevel surgery in order to correct deformities in the sagittal plane distal to the hip. Both clinical and instrument gait analysis results were recorded preoperatively, at one year (short term) and at five years (mid term) postoperatively.

Results: Clinically measured hip and knee movement improved at both short and mid term follow up. Correlations of clinically measured maximum hip and knee extension were significant at all three time points. Angles at terminal stance/toe off for hip and knee from kinematic data also showed significant correlations at all three time points.

Conclusions: Our study demonstrates that the hip flexion deformities encountered in these patients will improve spontaneously when the distal fixed knee flexion deformity is surgically corrected. Therefore correction at the knee allows the ground reaction force to assume a more normal position resulting in correction at the hip over time. This then removes the need for surgery at the hip level. This fact is especially important when applied to psoas lengthening as this procedure can cause significant reduction in propulsion power.

Key words: Cerebral palsy, Hip flexion deformity, Knee flexion deformity, Surgical treatment

Introduction

The mobility of children with bilateral spastic cerebral palsy (BSCP) deteriorates with time without intervention, especially during the pubertal growth spurt (1-4). In a natural history study, temporal gait parameters and gait kinematics were found to have deteriorated over time in 28 children with cerebral palsy (CP), including 19 patients with spastic diplegia, seven with hemiplegia, and two with quadriplegia (3).

The orthopaedic treatment of CP children includes conservative management and operative intervention. The surgical treatment in general consists of correcting bony deformities and muscle lengths. In order to avoid repetitive rehabilitations and compensatory problems secondary to longstanding deformities, several surgical interventions are combined in one session, known as single event multilevel surgery (SEMLS). The main goal of SEMLS is to achieve sagittal plane balance. This approach may be particularly important to improve gait in children with BSCP (5-9). A favourable outcome after multilevel surgery has been described for independent (10-12) and assisted walkers (13, 14). Instrumented clinical gait analysis provides essential biomechanical data to aid in the treatment decision making and to determine postoperative results in children with BSCP (15).

Progressive hip flexion deformity (HFD) is a common problem in ambulatory children with BSCP (16). The iliopsoas muscle is generally thought of as the major deforming factor in the development of static and dynamic HFD. The efficacy of psoas surgery for the treatment of hip flexor dysfunction in CP has long been the subject of debate (17, 18). Iliopsoas lengthening can be accomplished at different locations along the course of the tendon including lengthening above the pelvic brim (19), at the pelvic brim (20) or through complete tenotomy at the lesser trochanter with or without reattachment to the capsule (21). Intramuscular psoas lengthening has been reported as useful in reducing pelvic range of motion in the sagittal plane (22) and as well a safe and effective way to improve hip function of independently ambulatory children with CP while maintaining hip flexor power (17, 23). In contrast, Zwick et al (18) reported that patients who underwent psoas lengthening showed an increased anterior pelvic tilt and tendency towards genu recurvatum.

We wanted to evaluate the short- and mid-term outcome of surgical correction of fixed knee flexion deformity (KFD) on hip deformity in children with BSCP using the SEMLS approach. Our hypothesis was: Psoas contracture is not the primary cause for HFD, and for this reason HFD improves without psoas-lengthening after surgical correction of fixed knee flexion deformity at short and mid-term follow up.

Materials and methods

Study design and inclusion criteria

A retrospective study to evaluate the short- and mid-term outcomes after surgical correction of fixed KFD using the SEMLS approach in children and adolescents with BSCP was carried out. Inclusion criteria were a confirmed diagnosis of BSCP, Gross Motor Function Classification System (24) (GMFCS) level I, II or III and age 6-18 years. Prior injection of botulinum toxin A was allowed, as long as six months had elapsed since the last injection.

Eligible subjects had to have had a pre-operative gait analysis and preferably both a short-term and mid-term follow-up. Exclusion criteria were a diagnosis other than cerebral palsy, dystonic or mixed movement disorder, and patients who had had psoas lengthening surgery.

A total of 12 patients, 4 girls and 8 boys, with BSCP fulfilled the eligibility requirements. The mean age was 12.7 years (median 12.5, range 7-18) at the time of surgery. One patient was GMFCS level I, 9 patients were GMFCS level II, and two patients were GMFCS level III. The mean number of surgical interventions was 6.2 (median: 6.0, range: 4-10).

Clinical and gait evaluation

All participants had preoperative and at least one postoperative (short term 1.8 years, mid-term 5 years) 3D gait analysis including a thorough and standardised clinical assessment. The examination in the gait laboratory was performed by a physiotherapist and a human movement scientist trained in gait analysis. The clinical assessment included the examination of passive range of motion (ROM) with a goniometer. Clinical data (in degrees°) were collected for maximal hip extension, knee extension and ankle extension preoperatively and at short-term (mean follow-up of 1.8 years) and mid-term (mean follow-up of 5.0 years).

Instrumented gait analysis included kinematics, kinetics and dynamic electromyography (EMG) data, using a motion capture system (six camera VICON 460 system, Oxford Metrics Ltd., UK), two force plates (Kistler Instruments AG, Winterthur, Switzerland) and an eight channel surface EMG system (Zebris, Tübingen, Germany) (25). The patients walked at a self-selected speed. The Helen Hayes Marker set (26) was used and at least six trials were recorded. Anthropometric data were collected for appropriate scaling. All data were expressed as a percentage of the gait cycle using the Polygon software (Oxford Metrics Ltd., Oxford, UK). All data were up-loaded into Gaita Base (27). From Gaita Base hip angles, knee angles and ankle angles at terminal stance/toe off were downloaded.

Surgery

Surgical indications were based on a comprehensive biomechanical and clinical assessment including instrumented gait analysis and radiological evaluation. In this study SEMLS was defined as at least one surgical procedure, performed on two different anatomic levels (hip, knee, or ankle) on both sides of the body. The surgical procedures were not necessarily symmetrical and were not uniform across the group of patients but individually tailored to the child's needs. None of the patients had any surgical intervention to the psoas muscle, because this surgery is not carried out as a matter of routine in our institution. All operations were carried out under general anaesthesia. Post-operatively epidural infusions were established for pain control. All operative procedures were done or supervised by the senior author (RB).

Consent and ethical approval

All patients/parents gave written consent for surgery and for research purposes in accordance with local ethical committee requirements. The study was performed according to the declaration of Helsinki (World Medical Association).

Statistical analysis

Data were expressed as means with standard deviation. Normality of data was checked for using normal probability plots with application of the Shapiro Wilk test for normality. Correlations between range of motion at different anatomical levels (i.e. hip, knee, ankle) from both clinical and gait analysis data were compared at each time point using the Pearson correlation coefficient in parametric data and the Spearman Rank correlation in non-parametric data. The *t* test for dependent samples was used to compare between groups. The level of significance was set at $P = 0.05$ for all statistical tests. Statistical analysis was performed using the Statistica V6 software package (Statsoft Inc, 16 Tulsa, Oklahoma, USA).

Results

Clinical data

Clinical data as recorded for the total cohort preoperatively and at short and mid-term follow up are displayed in Table I. Preoperative clinical maximum hip extension showed a highly significant correlation to maximum knee extension ($p = 0.0022$). Similarly, preoperative clinical maximum knee extension versus maximum ankle extension showed a highly significant correlation (p = 0.0008). At short-term follow up, clinical maximum hip extension continued to correlate to maximum knee extension (p = 0.0291). Clinical maximum knee extension versus maximum ankle extension at short-term follow up was not significant (p = 0.1751). At mid term follow up clini-

Table 1. Clinical data of hip, knee, and ankle extension.

Pre SEMLS	N	Mean	Standard deviation	Minimum	Maximum
Clinical Hip Extension	24	-5.83	9.05	-30	10
Clinical Knee Extension	24	-6.25	12.53	-30	10
Clinical Ankle Extension	24	-0.21	12.02	-30	20
Short term follow up					
Clinical Hip Extension	24	0.83	6.70	-15	10
Clinical Knee Extension	24	-2.50	6.92	-20	10
Clinical Ankle Extension	24	3.54	9.61	-15	20
Mid term follow up					
Clinical Hip Extension	24	6.25	6.47	-10	15
Clinical Knee Extension	24	1.88	5.28	-10	10
Clinical Ankle Extension	24	2.92	11.51	-20	15

Clinical results (N = legs).
Negative values mean lack of extension.

cal maximum hip extension was still correlating significantly to maximum knee extension (p = 0.0324). Clinical maximum knee extension versus maximum ankle extension at mid term follow up was again highly significant (p = 0.0012). Therefore, this data revealed a statistically significant correlation of clinically measured maximum hip extension versus maximum knee extension at all three examinations points (preoperative, short and mid term follow up). The correlation results from the clinical data are summarised in Table II.

A longitudinal comparison of the clinical data for hip movement comparing the clinical values at all three time points (pre SEMLS, short and mid term) was carried out and showed:

- statistically significant difference between pre SEMLS and short term (p = 0.0034)
- statistically significant difference between pre SEMLS and mid term (p = 0.000036)
- statistically significant difference between short term and mid term (p = 0.0036)

Longitudinal comparison of the clinical data for knee movement showed:

- no significant difference between pre SEMLS and short term (but clinically improved by 3.75˚)
- statistically significant difference between pre SEMLS and mid term (p = 0.028)
- statistically significant difference between short term and mid term (p = 0.039)

Longitudinal comparison of ankle movements did not show any significant difference between the ankle values at pre SEMLS, short term and mid term follow up.

Table 2. Summary of correlation results of clinical findings.

	Preoperative	Short term	Mid term
Hip:Knee	R = 0.5942 P = 0.0022*	R = 0.4455 P = 0.0291*	R = 0.4378 P = 0.0324*
Hip:Ankle	R = 0.3281 P = 0.1176	R = 0.3741 P = 0.0717	R = 0.0949 P = 0.6591
Knee:Ankle	R = 0.6403 P = 0.0008*	R = 0.2862 P = 0.1751	R = 0.6219 P = 0.0012*

*P<0.05, statistically significant.

3D results from gait analysis

3D data were collected preoperatively for all 12 patients. One patient missed the first postoperative gait analysis, giving 11 patients with short-term results at a mean follow-up of 1.8 years (range 1-3). For 10 patients mid-term results were collected with a mean follow-up of 5.0 years (range 4-6). Two patients had not yet reached the time interval for mid-term follow-up gait analysis study but had undergone a standard clinical assessment, which did not indicate a deterioration clinically.

Figure 1 illustrates the sagittal plane kinematics of the entire cohort throughout the gait cycle, demonstrating the differences in these kinematic profiles at each different time point. Kinematic data at terminal stance for pelvis, hip, knee, and ankle angles as recorded for the total cohort preoperatively and at short and mid-term follow up is displayed in Table III. Preoperative sagittal kinematic data for knee versus pelvis angle showed a highly significant correlation (p = 0.0001). The same was shown for hip versus knee angle (p = 0.0312). Preoperative sagittal kinematic data for hip versus ankle angles showed no significant correlation (p = 0.0625).

At short term follow up, sagittal kinematic data at short term for knee versus pelvis angles showed no significant correlation (p = 0.1778). However, sagittal kinematic data at short term follow up for hip versus knee angles again showed a highly significant correlation (p = 0.0018). Sagittal kinematic data at short term for hip versus ankle angles showed a significant correlation (p = 0.0315).

At mid term follow up, sagittal kinematic data for knee versus pelvis angles showed a significant correlation (p = 0.0411) as pre-operatively. Sagittal kinematic data at mid term for hip versus knee angles continued to show a highly significant correlation (p = 0.0005). Sagittal kinematic data at mid term for hip versus ankle angles showed a significant correlation (p = 0.0105) as at short term follow up. Therefore a statistically significant correlation for sagittal kinematic data for hip versus knee angles was found at all three examinations (preoperative, short and mid term follow up). The correlation results from the kinematic data are summarised in Table IV. Figure 2 shows examples of the correlation curves demonstrating correlations between kinematic data at hip and knee at all 3 time points.

When a longitudinal comparison of the kinematic data for pelvic hip and knee angles was carried out, no significant difference in the angle at terminal stance was shown between any of the time points (pre SEMLS, short and mid term).

Table 3. 3d results from gait analysis (n= legs).

	n	Mean pelvis angle	Standard deviation pelvis angle	Mean hip angle	Standard deviation hip angle	Mean knee angle	Standard deviation knee angle	Mean ankle angle	Standard deviation ankle angle
Pre sagittal	24	15.57	9.25	22.25	11.84	53.07	14.72	-23.15	21.19
Short sagittal	22	19.88	6.91	19.99	11.73	48.62	12.64	-5.76	7.28
Mid sagittal	20	20.46	5.22	17.54	9.31	45.59	11.02	-9.33	9.8

All values at terminal stance of the gait cycle.

Table 4. Summary of correlation results of kinematic findings.

	Preoperative	Short term	Mid term
Pelvis:Hip	R = 0.1045 P = 0.6270	R = 0.4438 P = 0.0386*	R = 0.0623 P = 0.8060
Pelvis:Knee	R = -0.7087 P = 0.0001*	R = -0.2981 P = 0.1778	R = -0.4856 P = 0.0411*
Pelvis:Ankle	R = -0.3242 P = 0.1223	R = 0.0756 P = 0.7382	R =0.2512 P = 0.3146
Hip:Knee	R = 0.4405 P = 0.0312*	R = 0.6268 P = 0.0018*	R = 0.7337 P = 0.0005*
Hip:Ankle	R = -0.3860 P = 0.0625	R = 0.4595 P = 0.0315*	R = 0.5864 P = 0.0105*
Knee:Ankle	R = 0.2093 P = 0.3263	R = 0.5696 P = 0.0056*	R = 0.5351 P = 0.0221*

*P<0.05, statistically significant.

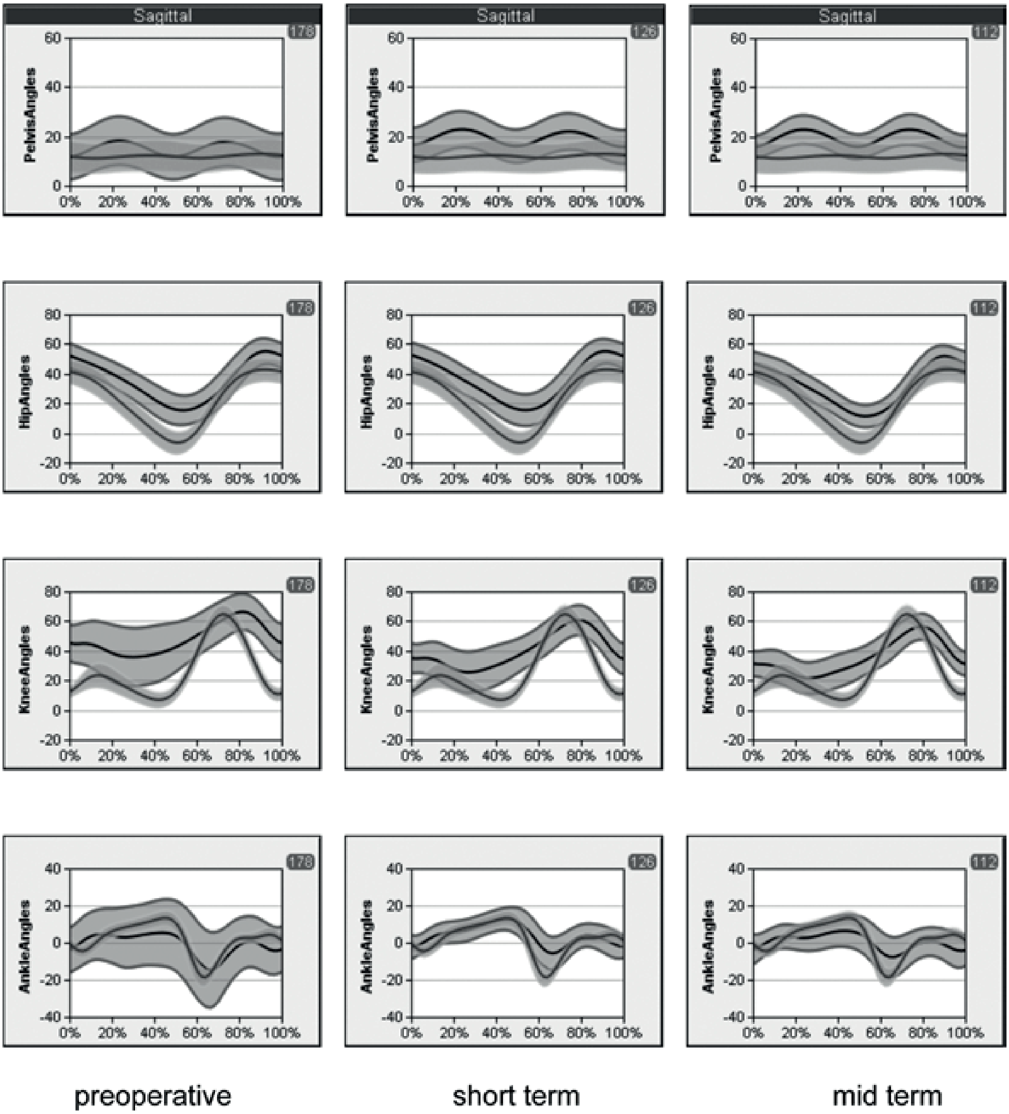

Fig. 1. *Sagittal plane kinematics. Narrow bands illustrate normal patients without gait pathology. Wide bands show all trials of the cohort (numbers at the top right indicate the number of recorded trials).*

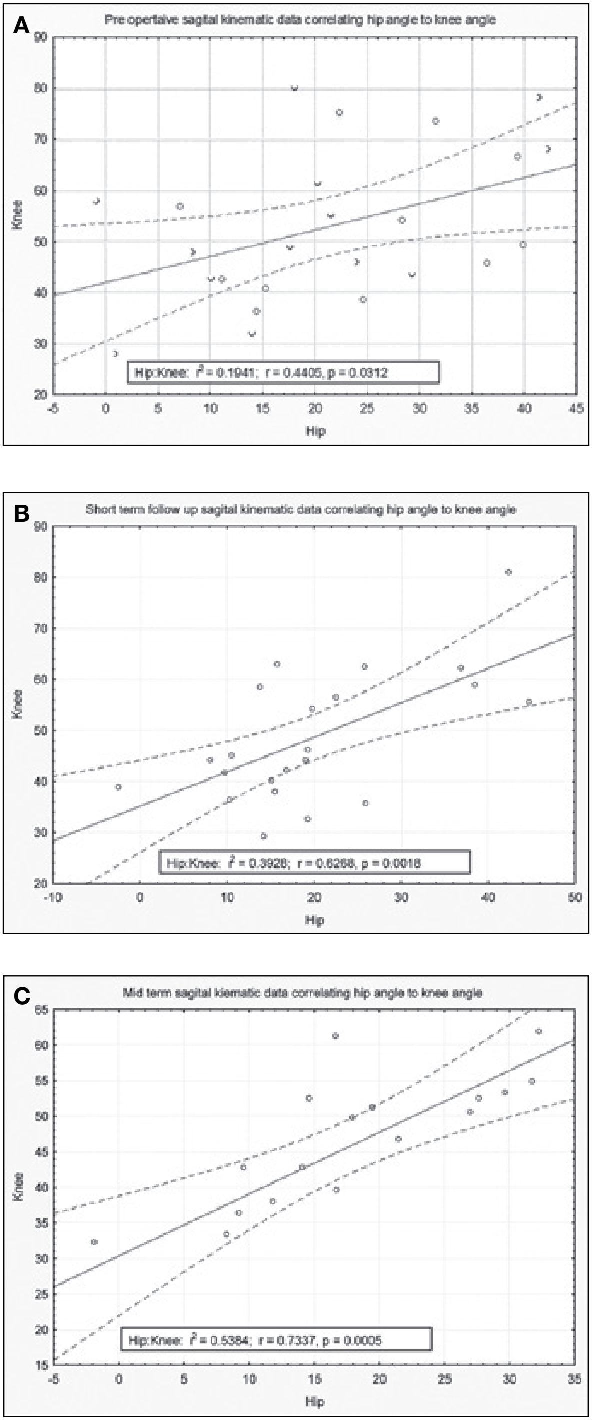

Fig. 2. Correlations of the 3D data (gait analysis) hip versus knee. A) preoperative; B) short term; C) mid term.

Longitudinal analysis of the kinematic data showed for ankle angles revealed a significant difference between pre SEMLS and short term (p = 0.0029), a significant difference between pre SEMLS and mid term (p = 0.027) and no significant difference between short and mid term (p = 0.384).

Statistical analysis of cadence, walking speed, stride and step length did not show any significant difference between pre SEMLS and short term, short,- and mid term values, and pre SEMLS and mid term results.

Discussion

The clinical results and the kinematic data from 3D-gait analysis show an improvement of sagittal angles by SEMLS at knee and hip joint although only the knee was corrected. Both the clinical and kinematic data demonstrate a significant correlation between knee and hip extension preoperatively and also at short and mid term follow up. This supports the hypothesis of this study that the hip flexion deformity is a secondary compensation in order to control knee extension. If the latter is improved the need for compensation at the hip resolves spontaneously and so does the hip deformity without psoas-lengthening.

Creating an external extensor moment at the knee by shifting the centre of mass (CoM) forward is the most plausible mechanism. In this case gravity needs to be considered which pulls the CoM down and thus flexes the hip. In this situation it is the hip extensors and not the hip flexors which need to hold the posture. Hip flexor activity in contrast would be deleterious as the muscles would pull the trunk to the floor as synergists to gravity. The constant need for forward bending results thus in secondary hip flexion deformity.

The present data showed that as the knee was able to extend more, so did the hip. The values for both joints improved following SEMLS procedures. A hip flexor lengthening can be avoided and thus weakening of these important muscles for increasing speed and stair climbing, unless there is an extreme and longstanding deformity. The mean number of surgical interventions per SEMLS session in our study was 6.2. Only the number of necessary were done as determined preoperatively by gait analysis and the number is unlikely to affect the results as the number of procedures was proportional to the severity of deformity in each patient with the aim of getting them all to the same level.

The hip: ankle correlations for clinical data did not show any significant difference at baseline, short,- and mid- term. Regarding the knee: ankle correlations we found statistically significant differences at baseline and mid term (Tab. II). The mean pelvic angles from the kinematics improved with time (from 15.6° at baseline to 20.5° at mid term). The mean ankle angles improved from severe equinus (-23.2°) to mid equinus (-5.8°, pre SEMLS to short term) and then silightly deteriorated at mid term (-9.3°, Tab. III).

Muscle–tendon lengthening surgery always carries the danger of muscle weakness (28). Over lengthening and subsequent muscle weakness should be avoided, especially as repairs of failed muscle-tendon lengthenings are difficult or even impossible. There are few published reports of the results of psoas lengthening (16-23, 29, 30) and there is still debate whether this procedure is effective. The psoas muscle is essential for powering hip flexion especially in swing, which is essential for propulsion during normal walking and to climb stairs. Basset et al (31) showed that lengthening of the psoas tendon during open reduction of dysplastic hips is associated with considerable atrophy of the psoas muscle. Those individuals with normal muscle innervation have, however, more possibility to compensate for the lack of a specific muscle than patients who already walk at their limits. For this reason we suggest avoiding surgery on the psoas muscle unless a deformity persists over time.

Our study has a number of limitations, which include small sample size, retrospective data analysis, lack of a control group and incomplete follow-up for some patients. The flexion contractures at hip, knee, and ankle are quite small, but with a large standard deviation. All data were collected carefully in a standardised protocol.

Conclusion

HFD depends significantly on knee extension. It deteriorates with increasing knee flexion and improves with decreasing knee flexion. This is because the CoM is shifted forward in order to create an external knee extension moment and thus to reduce knee extensor load. HFD resolves spontaneously and surgical lengtheining of the hip flexors in walking patients (according to our observations) is not necessary.

Financial support

This study was financially supported by the Swiss Society for Orthopaedic and Trauma Surgery (SGO).

Conflict of interest statement

There are no conflicts of interest.

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6

Chapter 7

Stability of Gross Motor Function Classification System after single-event multilevel surgey in children with cerebral palsy

Erich Rutz,Oren Tirosh, Pam Thomason, Alexej Barg and H Kerr Graham

Abstract

AIM There are conflicting reports about the stability of the Gross Motor Function Classification System (GMFCS) in children with cerebral palsy (CP) after orthopaedic surgery. We studied the stability of the GMFCS in children with bilateral spastic CP after single-event multilevel surgery, using the Gait Profile Score (GPS) as the primary outcome measure.

METHOD This was a retrospective cohort study of 107 children (46 females, 61 males) with bilat-

eral spastic CP, classified as GMFCS level II or III, who underwent surgery at a single tertiary institution between 1997 and 2008. The mean age at surgery was 10 years 7 months (SD 2y 8mo). The primary outcome measure was the GPS. Changes in GMFCS level were studied at multiple time points before and after intervention.

RESULTS Gait dysfunction was partially corrected, with a mean improvement of 28% in the GPS.

The GMFCS remained stable and unchanged in 95% of children and improved by one level in 5% of children. The improvement in GPS was three times the minimal clinically important difference. The mean age at final postoperative GPS assessment was 11 years 10 months (SD 2y 10mo) and at final GMFCS assessment was 15 years 7 months (SD 3y 9mo).

INTERPRETATION Stability of the GMFCS was confirmed in the majority of children with bilateral spastic CP after single-event multilevel surgery, despite statistically and clinically significant improvements in gait dysfunction and functional mobility. This information is important in realistic goal-setting and in counselling families.

What this paper adds

- Single-event multilevel surgery (SEMLS) significantly improves gait in children with spastic diplegia.
- GMFCS level remains stable in the majority of children after SEMLS.
- Changes in gross motor function are generally small.
- In our centre deterioration is unlikely and 5% of the children may improve by one GMFCS level.

Introduction

The Gross Motor Function Classification System (GMFCS) is a standardized method to classify gross motor function in children with cerebral palsy (CP) from 1 to 18 years of age.[1,2] It is a five-level categorical grading system which classifies differences in gross motor function that are meaningful and significant to children with CP and their families.[3] Distinctions between GMFCS levels are based on functional limitations with an emphasis on sitting and walking and the need for assistive devices and wheeled mobility. Children may be quickly and easily classified at any given time, such as during a clinic appointment or gait laboratory assessment, by determining which level best corresponds to the child's current gross motor function. A set of age-appropriate descriptive criteria are used and good correlations have been demonstrated between classifications assigned by parents and by health professionals.[1,2,4] Since the original description by Palisano et al.[1] in 1997, the GMFCS has become widely accepted, widely used, and is considered by many to be an essential tool to communicate about gross motor function in CP.[5,6]

The GMFCS is a valid and reliable classification tool which is expected to be relatively stable over time, with or without intervention. Palisano et al.[3] reported that 73% of 610 children with CP remained at the same GMFCS level at multiple ratings over time, and McCormick et al.[7] found that GMFCS level at age 12 years was highly predictive of adult gross motor function. Harvey et al.[8] reported that GMFCS level remained stable in the majority of ambulant children who had single-event multilevel surgery (SEMLS), even though improvements in mobility were found using the Functional Mobility Scale (FMS).

SEMLS is a programme of multiple orthopaedic procedures conducted during a single operative intervention and requiring a single hospital admission and period of rehabilitation.[9,10] The goal of SEMLS is to improve gait and functioning by correction of fixed musculoskeletal deformities in ambulant children with CP. The operative procedures consist of the correction of fixed contractures by muscle recessions and tendon lengthening, the correction of muscle imbalance by tendon transfers, and the correction of torsional bony deformities by rotational osteotomies of long bones and joint stabilization procedures.[9,10] Robust evidence for improvement in gait parameters after SEMLS has been reported in a number of controlled studies and in one randomized controlled trial.[11–13] The randomized controlled trial reported a mean improvement in Gross Motor Function Measure (GMFM) 66 of 4.9% at 24 months after SEMLS and that GMFCS levels remained stable in the majority of children.[13] However, other studies using the GMFCS as an outcome measure after orthopaedic surgery reported that the majority of the study participants improved by one level and that some participants improved by two or three levels.[14–16] In one of the studies, GMFCS levels were assigned retrospectively by chart review. To date there have been few intervention studies which have reported *prospectively* assigned GMFCS levels and objective measures of gait and function. The purpose of this study was to examine the stability of the GMFCS in a large group of children with CP, before and after SEMLS. The detailed surgical and gait outcomes are outside the scope of this study and will be reported elsewhere.

Method

Ethical approval for this study was granted under the audit provisions of the Ethics in Human Research Committee of The Royal Children's Hospital, Melbourne, Australia.

Inclusion criteria for this retrospective, cohort study were a confirmed diagnosis of CP with registration on the Statewide CP Register in Australia, a predominantly spastic movement disorder, bilateral involvement, GMFCS level II or III, age 6–18 years, and SEMLS between May 1997 and December 2008. In total, 107 children with bilateral spastic CP were eligible for inclusion. In addition, eligible participants were required to have undergone a preoperative gait analysis and at least one postoperative follow-up gait study, to provide objective measurement of changes in gait. Exclusion criteria were a diagnosis other than CP, dystonic or mixed movement disorder, age outside the range, and GMFCS level other than II or III.

Before SEMLS, each participant underwent a comprehensive assessment in the gait laboratory, which included recording of demographic and clinical data, assessment of GMFCS level, a standardized physical examination, radiological assessment, and three-dimensional gait analysis.[17] GMFCS level was recorded by an experienced physiotherapist, using age-appropriate descriptors.[1,2] Quantitative three-dimensional gait data were collected using a 50 Hz, six-camera Vicon 370 system (Oxford Metrics, Oxford, UK). Reflective markers were applied to the bony landmarks using a standardized procedure. Kinematic data were calculated using Plugin Gait (Oxford Metrics, Oxford, UK). The Gait Profile Score (GPS) was used to assess changes in gait function after SEMLS. The GPS was calculated from the root mean square difference in gait kinematics for an individual trial and the average kinematic data from children with no gait pathology.[18] Units were recorded in degrees and the larger the GPS, the more abnormal the participant's gait. The GPS was calculated for both legs on at least four individual gait cycles. The median GPS was calculated for each child using Gaitabase, a web-interfaced repository for gait analysis data.[19]

The indications for SEMLS were the presence of fixed musculoskeletal deformities that were considered to be adversely affecting gait and function.[20,21] Operative procedures for each SEMLS prescription were selected by evaluation of the above data, in discussion with the gait laboratory team and the child and parents or carers.[13,17] The selection and conduct of the operative procedures, postoperative care, and rehabilitation have been described in detail elsewhere.[13] During the first 12 months after SEMLS each child was reviewed in the gait laboratory every 3 months to assess progress in rehabilitation. On each of these occasions the gait was videotaped and GMFCS level and FMS were recorded. The FMS reports the level of assistance required to ambulate over distances of 5m, 50m, and 500m.[22] It is complementary to the GMFCS, is sensitive to change after SEMLS, and can be used as an outcome measure.[8] At 12 months after SEMLS, a full assessment was performed, including three-dimensional gait analysis and recording of GMFCS level and FMS. Children were then reviewed in the gait laboratory every 6 to 12 months according to clinical indications. GMFCS level and FMS were recorded at each gait laboratory assessment. All data sets were complete.

Statistical analysis

A Kolmogorov–Smirnov normality test was performed to verify that the data met the assumptions of a parametric test. A two–tailed paired-sample *t*-test was used for continuous variables. The level of significance (two-tailed) was set at a $p<0.05$. All statistical analyses were performed using SPSS version 20.0 (SPSS Inc., Chicago, IL, USA) and SigmaPlot version (Systat Software Inc., San Jose, CA, USA).

Results

Forty-six females and 61 males participated in the study, with a mean age at the time of SEMLS of 10 years 7 months (SD 2y 8mo; range 6y–17y). The mean interval between the first gait analysis and SEMLS was 7.5 months (SD 6.2mo; range 0–33mo) and between surgery and the first postoperative gait analysis was 15.6 months (SD 10.4mo; range 6–72mo). The mean clinical follow-up, including assignment of GMFCS level by an experienced physiotherapist, was 5 years (SD 2 years 8 months; range 2–12y). The median number of surgical procedures per SEMLS session was eight (range 4–14). The mean GPS decreased significantly from 15.4° (SD 3.9°; range 8.9–25.9°) to 11.1° (SD 2.5°; range 6.1–18.8°) ($p<0.001$).

The improvement in GPS was three times the minimal clinically important difference of 1.4°.[23] Despite clinically significant improvements in gait, GMFCS level was unchanged in 102 patients (95%) and changed in five patients (5%). The changes in

GMFCS were all to a higher level; three patients improved from GMFCS level III to II and two patients improved from GMFCS level II to I. Importantly, no patient deteriorated by a GMFCS level during the study period. In all but one child the change to the next higher GMFCS level occurred during the first year after SEMLS. Table I presents the characteristics of the cohort, Table II outlines the GMFCS levels, and Table III indicates the types of surgical interventions. See Appendix I for examples of specific patient case histories post SEMLS, including stable GMFCS level and change in GMFCS level.

Table 1. Summary of characteristics of the study cohort (n=107).

Characteristics	
Sex	46 females (43%), 61 males (57%)
Age at surgery	10y 7mo (SD 2y 8mo; range 6–17y; 25th–75th centiles 8–13y)
Time between preoperative three-dimensional gait analysis and SEMLS	7.5mo (SD 6.2mo; range 0–33mo; 25th–75th centiles 3–11mo)
Time between surgery and postoperative three-dimensional gait analysis	1y 4mo (SD 11mo; range 6mo–6y, 25th–75th centiles 1–1y)
Number of surgical procedures per SEMLS session	Eight procedures (range 4–14 procedures; 25th–75th centiles 6–9 procedures)
Age at postoperative assessment	11y 11mo (SD 2y 10mo; range 7–22y; 25th–75th centiles 9–15y)
Clinical follow-up	5y (SD 2y 8mo; range 2–12y; 25th–75th centiles 2–7y)
GPS preoperative	15.4° (SD 3.9°; range 8.9°–25.9°; 25th–75th centiles 12.3°–18.5°)
GPS postoperative	11.1° (SD 2.5°; range 6.1°–18.8°; 25th–75th centiles 9.1°–12.7°)
Change in GPS	4.3° (SD 3.7°; p<0.001a)
Change in GPS in relation to minimal clinically important difference	Threefold

aUsing a two-tailed paired sample t-test. SEMLS, single-event multilevel surgery; GPS, Gait Profile Score.

Table 2. Gross Motor Function Classification System (GMFCS) levels of the participants (*n*=107), *n* (%).

GMFCS level	Preoperative (mean age 10y 7mo; SD 2y 8mo)	Postoperative (mean age 15y 7mo; SD 3y 9mo)
I	0 (0)	2 (2)
II	74 (69)	75 (70)
III	33 (31)	30 (28)

Table 3. Types of surgical interventions performed.

	Right	Left	Total
FDO distal	14	12	26
FDO proximal	19	21	40
VDRO	37	38	75
VDRO extension	0	1	1
POTB (Gage)	17	17	34
PATB (Sutherland)	18	17	35
Unspecified PL	4	5	9
PALT	0	1	1
MHS	63	66	129
M&LHS	16	17	33
RF proximal release	3	3	6
RFT to gracilis	3	3	6
RFT to semitendinosus	53	57	110
RFT to tensor fasciae latae	1	1	2
ST to adductors	1	1	2
ST to tubercule adductor	9	8	17
Open ADD longus	3	4	7
Percutaneous ADD	1	1	2
Percutaneous ADD longus	35	36	71
Open TAL	8	6	14
Strayer	64	64	128
Baker	1	2	3
Vulpius	3	1	4
OCL	13	12	25
SMO	16	9	25
STF	3	4	7
Hindfoot valgus osteotomy	8	9	17
Recession of tibialis posterior (Frost)	2	3	5
Split transfer (SPLATT)	1	2	3

In total, 837 surgical procedures were performed in 107 SEMLS sessions. The median number of surgical procedures per SEMLS session was eight (range 4–14). FDO, femoral derotation osteotomy; VDRO, varus derotation osteotomy; POTB, psoas lengthening over the brim; PATB, psoas lengthening at the brim; PL, psoas lengthening; PALT, psoas lengthening at the lesser trochanter; MHS, medial hamstrings lengthening; M&LHS, medial and lateral hamstrings lengthening; RF, rectus femoris; RFT, rectus femoris transfer; ST, semitendinosus transfer; ADD, adductor lengthening; TAL, tendon Achilles lengthening; OCL, os calcis lengthening; SMO, supramalleolar osteotomy; STF, subtalar fusion; SPLATT, split tibialis anterior tendon transfer.

Discussion

The current definitions of CP all emphasize that CP is a disorder of movement and posture, leading to motor impairment.[24] It is secondary to a fixed, unchanging cerebral lesion which is acquired during early development *in utero*, during birth, and within the first year of life.[25] In contrast to the fixed cerebral lesion, the musculoskeletal pathology associated with CP is frequently progressive and may cause deterioration in gait, gross motor function, mobility, and health-related quality of life.[24,26] Given that the cerebral lesion is not remediable at this time, considerable effort is expended to ameliorate the effects of both the movement disorder and the fixed musculoskeletal pathology. When deformities become severe and fixed, various forms of orthopaedic surgery may be helpful.[26–28] For the ambulant child, the standard of care is considered to be SEMLS. This is supported by a number of cohort studies, a systematic review, and one randomized controlled trial.[11–13,29] Changes after SEMLS may include large improvements in gait, especially when musculoskeletal deformities are severe and underlying strength and selective motor control are relatively good. However, in a systematic review of SEMLS, changes in gross motor function were generally small and inconsistent.[29] This is in keeping with other interventions such as selective dorsal rhizotomy, which may result in large improvements in gait pattern and efficiency but small improvements in gross motor function.[30] Given that both neurosurgical procedures and orthopaedic surgery do not change the cerebral lesion, this is not surprising.

The GPS, recently published by Baker et al.,[18] is a summary statistic of gait that quantifies an individual's gait abnormalities relative to normative data. The improvement in GPS in this study was three times the minimum clinically important difference.[23] However, despite statistically and clinically significant improvements in gait dysfunction, GMFCS level remained stable in the majority of patients in our cohort.

The prevalence of crouch gait may be related to tendon Achilles lengthening procedures, which often result in loss of independent ambulation and a change down from GMFCS level II to GMFCS level III. Severe crouch gait is now at low levels (0–4%) in our population because of early non-operative management and avoidance of single-level gastroc-soleus lengthening.[31]

The GMFCS is a vital prognostic tool because it is stable over time.[3,7] However, recent outcome studies have reported improvement in GMFCS level in the majority of patients, by one, two or even three levels.[14–16,32] These reports include changes after selective dorsal rhizotomy and orthopaedic surgery.[33] However, most of these studies had methodological problems, including retrospective assignment of GMFCS level, sometimes from chart review, or a lack of supporting data from simultaneous measurements of either gait or gross motor function. In this study, we were able to demonstrate two main findings. First, gait function was improved in most patients after SEMLS and, second, the GMFCS level remained stable, over time, in most patients.

Following successful gait correction surgery, with documented improvements in overall gait and individual gait parameters, GMFCS level remains stable in the majority of children with CP. In this study, 5% of children improved by one GMFCS level and none deteriorated. In only a small number of children, who are preoperatively at an interface between GMFCS levels, will GMFCS level change after SEMLS. This confirms that the GMFCS is an ideal tool for classification of children with CP, establishing long-term gross motor prognosis and, by implication, goal-setting. The improvement in gait after SEMLS ranges between 20% and 50% in reported studies.[11–13,29] However, there is considerable variation in the mean change reported in most studies, indicating that changes in gait after SEMLS are variable. The factors responsible for this variation are poorly understood.

Gait improvements of this magnitude (summarized in a systematic review[29]) are clinically significant and would be easily recognized by parents watching their child walk. They are also easily detected using the FMS by a reduction in the need for assistive devices.[8] Reported changes in gross motor function after SEMLS vary from 0% to 4.9%, again with a significant standard deviation about the mean.[11,13,29] Given the direct relationship between GMFM and GMFCS it is readily apparent that some changes in GMFCS level can be expected after interventions that result in a measurable change in GMFM. Equally, the magnitude of change in GMFM reported after both selective dorsal rhizotomy and SELMS would not be expected to result in improvements in GMFCS in the majority of patients.[13,30] Instead, children at the interface between GMFCS levels or those whose gait improves by one or two SDs above the mean change may achieve a higher GMFCS level.[13,34]

It may be that categorical measures are more prone to bias, especially when assigned retrospectively rather than as continuous measures, determined prospectively. In this study, GMFCS levels were collected prospectively by experienced physiotherapists with a close knowledge of the child's functional abilities, derived from multiple domains.[17] The expectation was that GMFCS would be stable and any bias in recording GMFCS levels may have been in the direction of stability rather than change.

Nevertheless, the modest upwards changes recorded in this population over time are in agreement with measured changes in gait (GPS), functional mobility (FMS), and GMFM reported by our group in other studies.[8,13,21] Clinicians and parents are in a constant quest for more effective interventions and better outcomes for children with CP. However, it is vitally important to be able to offer realistic counselling as to the likely long-term outcomes before invasive interventions. For SEMLS in our centre, this would include large improvements in gait, less need for assistive devices, small improvements in gross motor function, and stability of the GMFCS. Time will tell if biological reparative therapies, such as the use of stem cells, can achieve the breakthrough in changing GMFCS levels sought by parents and clinicians.

In summary, stability, rather than change, is to be expected in GMFCS levels after major interventions. The GMFCS is a categorical classification tool and should not be used as a primary outcome measure. When changes in GMFCS are reported after interventions, to summarize functional changes, the data should be supported by continuous measures such as gait scores and by direct measurement of gross motor function, using the GMFM.

Acknowledgements

The authors wish to thank the Swiss National Science Foundation for financially supporting this study, Associate Professor Susan Donath MA, BSc Statistician, Murdoch Childrens Research Institute for statistical advice, and Mary Sheedy and Dr Daniela Rutz, for their help in the preparation of this manuscript.

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APPENDIX I: EXAMPLES OF PATIENT CASE HISTORIES POST SEMLS

Example 1: Stable GMFCS after SEMLS, despite major gait improvements

A 9-year-old male with spastic diplegia related to preterm birth was assessed for SEMLS because of rapidly increasing lower limb contractures and deteriorating gait. The patient had severe jump gait and functioned at GMFCS level II, but could walk for limited distances only (FMS 5,5,1). His preoperative GPS was 18.5°, confirming severe gait dysfunction. At 12 months after SEMLS his GPS was 9°, a 50% improvement. His FMS was 6,6,5 indicating that he could walk independently in the community. His GMFCS remained at level

II. Summary: major improvement in gait and FMS, stable GMFCS.

Example 2: Change in GMFCS level after SEMLS – moving up to a new level.

A 9-year-old female with severe diplegia, obesity, and intellectual disability presented with gait deterioration. The patient was GMFCS level III, FMS C,2,1, and her GPS was 15.9°. At 12 months after SEMLS she was GMFCS level II, FMS 5,5,1, and her GPS was 13.5°. Her GMFM66 was 51.6 before surgery and 60.1 at 24 months after SEMLS. Improvements in gait and function have been maintained at 5-year follow-up, with stable GMFCS.

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Chapter 8

Are results after single-event multilevel surgery in cerebral palsy durable?

Erich Rutz, Richard Baker, Oren Tirosh and Reinald Brunner

Abstract

Background Gait and function may deteriorate with time in patients with spastic diplegia. Single-event multilevel surgery often is performed to either improve gait or prevent deterioration. However it is unclear whether the presumed gait improvements are durable.

Questions/purposes We therefore determined whether
(1) single-event multilevel surgery improves gait in patients with spastic diplegia and
(2) whether the improvedfunction is durable.

Methods We retrospectively reviewed the data of 14 patients with spastic diplegia. At the time of surgery, one patient had gross motor Level I function, 10 patients had Level II function, and three patients had Level III function. There were four females and 10 males with a mean age of 13 years (range, 7–18 years). The mean number of orthopaedic procedures per single-event multilevel surgery session was 7.4 ± 2.8 (median, 6.5; range, 4–15). We used instrumented gait analysis to determine joint ROM, movement analysis profiles, and the gait profile score. The minimum followup was 1 year (mean, 2 years; range, 1–3 years).

Results At last followup, movement analysis profiles for knee flexion, for ankle dorsiflexion, and for foot progression improved as did the gait profile score. Additional surgery after the index procedure was performed in nine of the 14 patients because of relapse of the original or new gait problems. Major surgical adverse events occurred in one of the 14 patients.

Conclusions Severe gait dysfunction in patients with spastic diplegia can be improved short-term in one operative session by single-event multilevel surgery, but to preserve the early improvements many patients require additional surgery. It is unknown whether the improvements will last for decades.

Level of evidence Level IV, therapeutic study. See Guidelines for Authors for a complete description of levels of evidence.

Introduction

Without intervention, gait and function in patients with spastic diplegia may deteriorate with time, especially during the pubertal growth spurt [4, 14, 22]. In one study, deterioration in gait parameters occurred in all 18 subjects after a mean followup of only 32 months [14]. In another natural history study, temporal gait parameters and gait kinematics deteriorated in all 28 patients with cerebral palsy, including 19 with spastic diplegia, seven with hemiplegia, and two with quadriplegia [4].

Management of gait dysfunction in patients with cerebral palsy may include nonoperative measures, such as physiotherapy [45], orthotics [5], casting [19], and injections of botulinum toxin type A [17]. Despite appropriate nonoperative management, however, the majority of patients with spastic diplegia will have fixed contractures and bony deformities develop which require surgical correction to maintain gait or even sitting functions. To achieve sagittal plane balance and avoid repeated episodes of surgery and rehabilitation, multiple surgical interventions may be combined in one session, known as single-event multilevel surgery [7, 23]. This approach may be particularly important to improve gait in patients with spastic diplegia [11, 12, 37]. Single-event multilevel surgery was first described by Norlin and Tkaczuk in 1985 [23] and Browne and McManus in 1987 [7]. In these studies the concept of single-event multilevel surgery was introduced and the advantages of one-session surgery such as reduction in the number of operations and, possibly, reduced chance of reoperation and complications are mentioned. Norlin and Tkaczuk reported a 5-year followup of the original cohort in 1992 [24]. They reassessed 23 subjects by means of video-analysis rather than three-dimensional (3-D) gait analysis. In their cohort none of the patients had recurrences of the deformities and they had normal growth-related development of their gaits. However, these studies predated the widespread availability of instrumented gait analysis and the development of valid functional scales. Using a combination of gait parameters and the physiologic cost index, Nene et al. [21] in 1993 reported the function of 18 patients who had single-event multilevel surgery. Improvements in a predominance of gait parameters after multilevel surgery were reported for independent [36, 40, 47] and assisted walkers [18, 46]. Thomason et al. [42] published a pilot report of the first randomized clinical trial of single-event multilevel surgery. In a subsequent report of thesame study [43], Thomason et al., reported statistically significant and clinically important improvements: at 12 months after surgery they noted absence of joint contractures, correct muscle lengths and lever arms, and gait kinematics. However, functional improvements, such as Gross Motor Function Measure (GMFM66), were not seen until 2 years after surgery. These improvements in gait and function were maintained at 5 years after surgery. The most comprehensive of recent studies reporting single-event multilevel surgery is a prospective, multicenter study with a control group, performed by Gorton et al. [10]. They included a group of outcome measures, including a summary statistic of gait (the Gillette gait index) [38], a measure of gross motor function (the Gross Motor Function Classification System or GMFCS) [26], a measure of health-related quality of life, and measurements using the Pediatric Outcomes Data Collection Instrument [10]. The authors showed on the basis of a matched concurrent data set of 75 patients with spastic cerebral palsy (age 4 to 18 years), an improvement in function after 1 year for a surgical group compared with a nonsurgical group.

Instrumented clinical gait analysis consists of various combinations of kinematic, kinetic, and EMG data and is interpreted with supporting clinical data. It provides essential biomechanical data for decision-making and objective outcome assessment [9]. The movement analysis profile and gait profile score [3] were developed to provide graphic and quantitative summaries of kinematic gait data, and a single summary statistic of gait. The individual movement analysis profile domains show which gait parameters have changed and by how much. The gait profile score provides a single measure to help determine if overall gait function has improved or deteriorated. The movement analysis profile domains highlight specific gait deviations but they have not replaced detailed data interpretations required for surgical decision-making [9]. The movement analysis profile describes the magnitude of deviation of nine kinematic variables over the gait cycle. The gait profile score [3] reduces all information to one number that reflects the quality of the gait pattern with respect to normality; it is a summary statistic of gait, which quantifies the subject's gait abnormalities relative to nor-mative data. A higher figure indicates a more abnormal gait pattern with respect to normality, and a lower value indicates an improvement toward normal gait function. Based on an analysis of the difference in median gait profile score for patients classified at different levels of the Functional Assessment Questionnaire [25], the minimal clinically important difference for the gait profile score is 1.6° [2].

The purposes of this study were to determine whether (1) single-event multilevel surgery improved gait in patients with spastic diplegia; and (2) whether the improved function is durable.

Patients and Methods

We retrospectively studied all 14 patients with cerebral palsy and severe gait dysfunction who underwent single-event multilevel surgery between September 2004 and June 2008. Inclusion criteria were a confirmed diagnosis of spastic diplegia, GMFCS Levels I, II, or III, and ages 6 to 18 years. Prior injection of botulinum toxin A was allowed, as long as 6 months had elapsed since the last injection. Eligible subjects had to have had a preoperative gait analysis and most had short-term (ie, 1 to 3 years; n = 13) and mid-term (ie, 4 to 6 years; n = 12) followup gait studies. Exclusion criteria were a diagnosis other than cerebral palsy, dystonic or mixed movement disorder, outside the age range, and GMFCS Levels IV and V. There were four females and 10 males included in the study. They had a mean age of 13 years (median, 12.5; range, 7–18 years) at the time of preoperative 3-D gait analysis. One patient had GMFCS Level I function, 10 had Level II function, and three had Level III function. One patient had previous surgical procedures (Table 1). The mean age of the patients at the time of single-event multilevel surgery was 14 years (median, 13 years; range, 8–18 years). No patients were lost to followup. The minimum followup was 1 year (mean, 1.8 years; range, 1–3 years). No patients were recalled specifically for this study; all data were obtained from medical records. The change in the gait profile score was 10°, and this change, in relation to the minimum clinically important difference, was seven times. All patients and/or parents gave written consent for participation in the study in accordance with local ethical committee requirements. The study was performed according to the declaration of Helsinki.

Table 1. Details of the 14 patients at the first 3-D gait analysis.

Gender	10 males, 4 females
Age (years)	Mean, 12.8 ± 3.30;
	Median, 12.5;
	Range, 7–18
GMFCS	
Level I function	1 patient
Level II function	10 patients
Level III function	3 patients
Previous surgery	1 patient

3-D = three-dimensional; GMFCS = Gross Motor Function Classification System.

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Three-dimensional data were collected preoperatively for all 14 patients. One patient (Patient 10) missed the first postoperative gait analysis leaving 13 patients with short-term data at a mean followup of 2 years (range, 1–3 years). For 12 patients followup data were collected with a mean followup of 5 years (range, 4–6 years). Two patients were followed not long enough to have mid-term followup gait studies (Patients 9 and 13). The examination in our gait laboratory was performed by a physiotherapist and a human movement scientist trained in gait analysis. The clinical assessment included examining passive ROM by using a goniometer, the spasticity assessment using the Modified Ashworth Scale Instructions [6] (scale, 0–4), and the manual muscle strength test [8, 20] (scale, 0–5) of the lower limb muscles. The instrumented gait analysis included kinematics, kinetics, and dynamic EMG data using a motion capture system (6-camera Vicon 460 system™, Oxford Metrics Ltd, Oxford, UK), two force plates (Kistler Instruments AG, Winterthur, Switzerland), and an eight-channel surface EMG system (Zebris[1], Tübingen, Germany). The patients walked at their self-selected speed. The Helen Hayes marker set [15] was used, and at least six trials were recorded. Anthropometric data were collected for appropriate scaling. Surface EMG readings were recorded simultaneously. Bipolar Ag/AgCl surface electrode pairs (electrode diameter 10-mm and interelectrode spacing of 22-mm) were placed bilaterally over the medial gastrocnemius, tibialis anterior, rectus femoris, and semitendinosus muscles. For electrode placement the Surface ElectroMyoGraphy for the Non-Invasive Assessment of Muscles (SENIAM) [13] recommendations for surface EMG were followed. The ground electrode was placed over the tibial tuberosity. The EMG signals were band-pass filtered (10–700 Hz) and collected at a sampling rate of 2500 Hz. All data were expressed as a percentage of gait cycle, using the Polygon software (Vicon Polygon, Oxford Metrics Ltd, Oxford, UK). From the 3-D gait data, temporal parameters (cadence, stride length, and walking speed), the movement analysis profile, and gait profile score were calculated [3]. All patients underwent at least one preoperative and one postoperative gait analysis. All data were uploaded into GaitaBase (http://gaitabase.rch. org.au) [44]. The mean interval between preoperative gait analysis and surgery was 0.8 years.

All patients had preoperative botulinum toxin test injections before muscle lengthening surgery [33]. Surgical indications were based on comprehensive biomechanical and clinical assessments, including an instrumented gait analysis and clinical and radiologic evaluations. The indications for single-event multilevel surgery were:
(1) documented deterioration in gait and function during the last 12 months, (2) fixed musculoskeletal deformities (contracture, torsion, joint instability), (3) evidence from the diagnostic matrix, including 3-D gait analysis, that deformity correction at two

different anatomic levels (the hip, knee, or ankle) on both sides of the body had a realistic chance of improving gait and function (Appendix 1). The contraindications were: (1) severe weakness, (2) uncontrolled spasticity or dystonia, (3) progressive neurologic disorder (eg, hereditary spastic paraparesis), (4) a patient who was unable to perform postoperative rehabilitation because of cognitive, behavioral, geographic, financial, or social factors. For the purposes of this study, single-event multilevel surgery was defined as at least one surgical procedure that was performed on two different anatomic levels (hip, knee, or ankle) on both sides of the body. The surgical procedure did not need to be symmetric and was not uniform, but individually tailored to the patient's needs. All operations were performed with the patient receiving general anesthesia. The mean number of orthopaedic procedures was seven (median, 6.5; range, 4–15 procedures). All operative procedures were performed or supervised by the junior (ER) or senior author (RB). Perioperative antibiotics were used and postoperative epidural infusions were administered to all the patients for pain control. All patients remained as inpatients for 5 to 10 days after surgery and were discharged wearing below-the-knee plaster casts (except in case of isolated tibialis posterior lengthenings) with knee immobilizers and use of appropriate assistive devices as indicated by their GMFCS level. The patients were first assessed 6 weeks postoperatively with radiographs to check healing of the surgical incisions and consolidation of the osteotomies and bony procedures. Custom-fitted ankle-foot orthoses were provided for all patients with surgery at the ankle level. After 6 weeks all patients followed an intensive rehabilitation program performed three to five times per week for 12 weeks or 4 to 6 weeks as inpatients in a rehabilitation center. The aim of this postoperative rehabilitation was to improve ROM, strength, balance, and function. After this time all patients had semiannual clinic visits.

Adverse events related to surgery were classified as mild if they resolved spontaneously, moderate if they resolved completely after simple treatment, or severe if there was a permanent deficit [42].

Paired data were assessed using the paired t-test and sequential data were assessed using repeated ANOVA with Bonferroni post hoc analysis (SPSS[1] software, Version 15.0; SSPS Inc, Chicago, IL, USA). We determined differences in walking speed, cadence, and stride length between preoperative and the two followup times using t-tests. We determined differences in each movement analysis profile and the gait profile score between the preoperative and postoperative assessments using ANOVA.

Table 2. Temporal parameters for baseline versus short-term post-operative gait.

Parameter	Preoperation	Short-term	p value
Walking speed (m/second)	0.83 ± 0.26	0.90 ± 0.27	0.3200
Cadence (steps/ minute × 100)	1.86 ± 0.34	1.83 ± 0.39	0.8059
Stride length (m)	0.88 ± 0.19	0.98 ± 0.20	0.0744
Movement analysis profile (in degrees)			
Pelvic tilt	9.30 ± 4.75	11.53 ± 8.14	0.2330
Pelvic obliquity	5.60 ± 3.40	4.61 ± 1.41	0.8388
Pelvic rotation	10.72 ± 5.91	8.01 ± 3.79	0.0549
Hip flexion	18.83 ± 10.12	17.98 ± 8.51	0.7456
Hip abduction	6.71 ± 2.67	6.13 ± 2.00	0.3818
ip rotation	15.60 ± 10.10	11.97 ± 5.15	0.1086
Knee flexion	29.28 ± 16.94	18.49 ± 7.52	0.0045*
Ankle dorsiflexion	13.27 ± 14.17	8.00 ± 2.74	0.0005*
Foot progression	29.34 ± 24.36	14.17 ± 6.97	0.0036*
Gait profile score (in degrees)	20.00 ± 8.67	12.92 ± 3.57	0.0003*

* Statistically significant.

Table 3. Temporal parameters for baseline to mid-term postoperative gait.

Parameter	Preoperation	Mid-term	p value
Walking speed (m/second)	0.72 ± 0.27	0.95 ± 0.23	0.0026*
Cadence (steps/ minute × 100))	1.73 ± 0.44	1.84 ± 0.24	0.3025
Stride length (m)	0.81 ± 0.20	1.03 ± 0.19	0.0003*
Movement analysis profile (in degrees)			
Pelvic tilt	8.70 ± 4.11	9.08 ± 3.67	0.7322
Pelvic obliquity	5.50 ± 3.51	4.06 ± 2.00	0.0881
Pelvic rotation	9.74 ± 5.65	8.84 ± 3.72	0.3116
Hip flexion	20.29 ± 10.20	13.03 ± 4.91	0.0029*
Hip abduction	7.02 ± 2.91	6.46 ± 2.09	0.4488
ip rotation	13.83 ± 8.98	11.36 ± 6.27	0.2740
Knee flexion	32.18 ± 16.54	15.88 ± 4.95	< 0.0001*
Ankle dorsiflexion	20.62 ± 14.44	9.08 ± 3.21	0.0003*
Foot progression	29.80 ± 25.28	12.90 ± 6.86	0.0027*
Gait profile score (in degrees)	20.70 ± 8.77	11.05 ± 2.62	< 0.0001*

* Statistically significant.

Table 4. Comparison of short- and mid-term parameters operative gait.

Parameter	Short-term	Mid-term	p value
Walking speed (m/second)	0.86 ± 0.27	0.97 ± 0.23	0.4720
Cadence (steps/ minute × 100))	1.80 ± 0.41	1.85 ± 0.24	0.9814
Stride length (m)	0.96 ± 0.20	1.05 ± 0.20	0.4316
Movement analysis profile (in degrees)			
Pelvic tilt	11.97 ± 8.80	9.38 ± 3.68	0.4719
Pelvic obliquity	4.35 ± 1.07	4.00 ± 2.07	0.8873
Pelvic rotation	7.55 ± 3.69	6.89 ± 3.89	0.9876
Hip flexion	18.78 ± 9.05	12.83 ± 5.07	0.0712
Hip abduction	6.04 ± 1.72	6.32 ± 2.12	0.8002
ip rotation	11.61 ± 5.33	10.83 ± 6.17	0.6951
Knee flexion	19.19 ± 7.90	15.34 ± 4.77	0.7872
Ankle dorsiflexion	8.19 ± 2.90	8.76 ± 2.93	0.8854
Foot progression	14.51 ± 7.36	12.20 ± 6.27	0.9873
Gait profile score (in degrees)	13.25 ± 3.78	10.77 ± 2.48	0.5099

Results

After single-event multilevel surgery there was an improvement in gait: the movement analysis profile for knee flexion, ankle dorsiflexion, and foot progression improved. The temporal parameters for baseline versus short-term postoperative gait (Table 2) are shown, and the temporal parameters for baseline versus mid-term postoperative gait were compared (Table 3). The parameters for short-term versus mid-term postoperative gait are shown (Table 4). The preoperative, short-term, and mid-term kinematics (Fig. 1), and movement analysis profile are shown (Fig. 2). The mean gait profile score improved from 20° to 13° (Table 2). The change was 7° and this change, in relation to the minimum clinically important difference, was five times. There was improvement in walking speed and stride length. The movement analysis profile for hip flexion, knee flexion, ankle dorsiflexion, and foot progression showed remarkable improvements (Table 3). The mean gait profile score improved from 21° to 11°. Nine patients (64%) had additional surgery after single-event multilevel surgery for further gait correction (Table 5) combined with plate removal (when indicated). New problems were found in five patients (Patient 1, hip flexion deformity; Patient 2, unilateral hip displacement; Patient 11, severe pes planovalgus; Patient 13, over-lengthening of the tibialis anterior tendon; Patient 14, short adductors) (Table 5), and one of the procedures needed revision because of relapses in four patients (Patients

3, 4, 7, and 8). Therefore no important changes were noted between short-term and mid-term followups (Table 4). The mean gait profile score at short-term was 13° and at mid-term 11°. The change in gait profile score was 2°, and this change, in relation to minimal clinically important difference, was two times.

One patient (Patient 4) had a deep infection develop after tendoachilles lengthening. The deep infection rate was 1% for all interventions. No other complications occurred and this was the only adverse event.

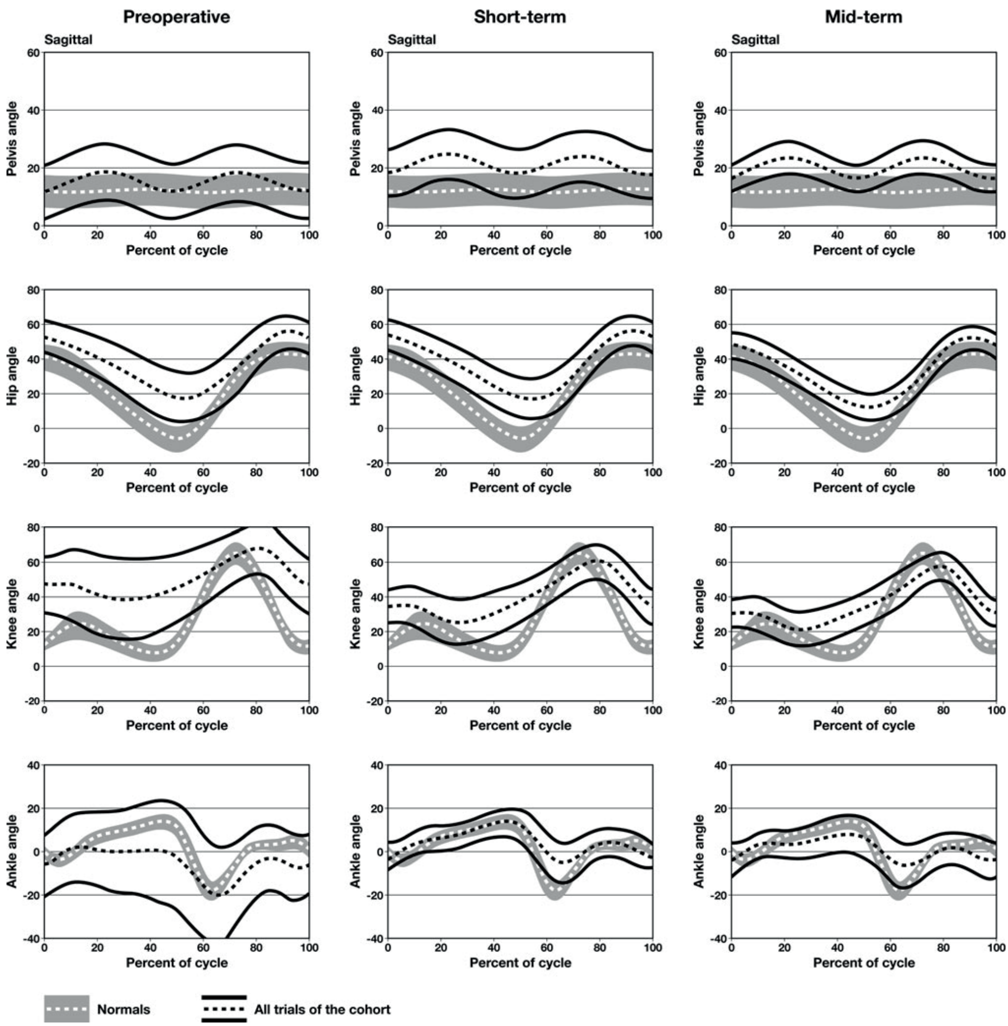

Fig. 1. These graphs illustrate the preoperative sagittal plane kinematics at the short-term and mid-term followups. The gray band is the normal reference range. The dotted line is the mean kinematic for the study group. The substantial improvements in the sagittal plane kinematic scan be seen; these are more clearly and quantitatively illustrated in Figure 2.

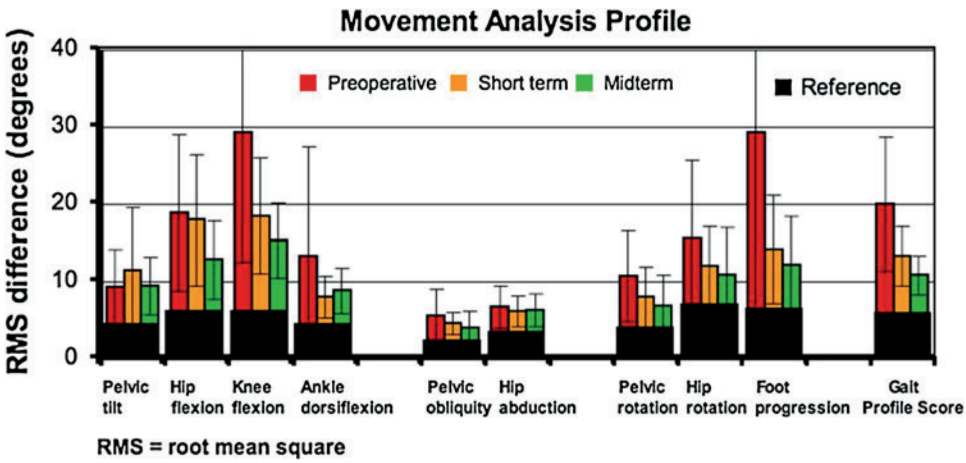

Fig. 2. The movement analysis profile and gait profile score are illustrated here at preoperative, short-term followup, and mid-term followups. Decreases in the movement analysis profile and gait profile scores are an improvement toward more normal gait.

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Discussion

The advantages of single-event multilevel surgery for gait dysfunction in patients with spastic diplegia include less time spent in the hospital and the necessity of a single period of rehabilitation. This may result in cost savings, but we are not aware of any cost benefit analyses. The first reports of single-event multilevel surgeries had a narrow focus on safety and feasibility [7, 23, 24]. Efficacy has been established in studies [10, 18, 21, 32, 34–36, 40, 43, 46, 47] which have included gait data and functional outcomes. Most studies have lacked coherent methods to determine if specific procedures have been effective and the effectiveness of the overall single-event multilevel surgery and rehabilitation package [7, 11, 21, 24, 36, 37]. The aim of our study was to investigate (1) whether single-event multilevel surgery improved gait in patients with spastic diplegia, and (2) whether the improved function is durable. A previous study by Norlin and Tkaczuk assessed 23 subjects at the 5-year followup to determine the long-term success of single-event surgery [24]. However, their study only included video recordings as a means of assessment, whereas our results are based on gait analysis. After 5 years they did not find any recurrences. In contrast, in our study 2/3 of the patients required additional surgery because of relapses or new biomechanical problems.

Our study has numerous limitations. First, we had a small sample size of 14 patients. The indications for single-event multilevel surgery are relatively uncommon; we did include all patients who had this sort of surgery. Second, we lacked control groups of

Table 5. Surgical details of all patients.

Patient number and gender	GMFCS level	Age at single-event multilevel surgery	Single-event multilevel surgery	Short-term followup (3-D gait analysis)	Additional surgery (after single-event multilevel surgery)	Mid-term followup (3-D gait analysis)
1 Male	III	16 years 2 months	Bilateral open adductor lengthening, bilateral medial hamstring lengthening, bilateral lateral hamstring lengthening, bilateral Baumann procedure, bilateral rectus release, left distal femoral derotation osteotomy, bilateral patellar tendon shortening, bilateral psoas lengthening	12 months	Bilateral proximal femoral extension osteotomy, bilateral psoas lengthening (3 years 1 month), new problem	4 years
2 Female	II	9 years 10 months	Bilateral open adductor lengthening, bilateral Baumann procedure	24 months	Right open hip reconstruction (3 years 5 months), new problem	6 years
3 Male	II	7 years 9 months	Left distal femoral derotation osteotomy, bilateral medial hamstring lengthening, bilateral Baumann procedure, left rectus release	12 months	Left medial hamstring lengthening (4 years 2 months), relapse	5 years
4 Female	II	10 years 11 months	Bilateral medial hamstring lengthening, bilateral lateral hamstring lengthening, bilateral tendoAchilles lengthening (2 months later, revision right tendoAchilles)	24 months	Bilateral distal femoral extension osteotomy, bilateral patellar tendon shortening, bilateral medial hamstring lengthening (2 years 7 months), relapse	4 years
5 Male	II	16 years 5 months	(2 years previous, bilateral varus derotation osteotomy) bilateral psoas lengthening, bilateral open adductor lengthening, bilateral medial hamstring lengthening	3 years	No additional surgery	6 years
6 Female	II	12 years 6 months	Bilateral Baumann procedure, bilateral medial hamstring lengthening	3 years	No additional surgery	4 years
7 Male	II	16 years 9 months	Bilateral patellar tendon shortening, bilateral distal femoral extension osteotomy, bilateral medial hamstring lengthening, right tendoAchilles shortening, bilateral subtalar fusion	12 months	Bilateral medial hamstring lengthening (3 years 5 months), relapse	4 years

8 Female	III	12 years 2 months	Bilateral open adductor lengthening, bilateral medial hamstring lengthening, bilateral patellar tendon shortening, right internal rotation supramalleolar osteotomy, right os calcis lengthening (Evans procedure)	3 years	Bilateral patellar tendon shortening, right tendoAchilles shortening (3 years 5 months), relapse	5 years
9 Male	II	10 years 8 months	Bilateral open adductor lengthening, bilateral medial hamstring lengthening, bilateral patellar tendon shortening, right distal femoral extension osteotomy, bilateral external rotation supramalleolar osteotomy	24 months	No additional surgery	None
10 Male	II	18 years 2 months	Bilateral open adductor lengthening, bilateral medial hamstring lengthening, bilateral rectus release, bilateral tendoAchilles lengthening, bilateral tibialis posterior lengthening	None	No additional surgery	6 years
11 Male	II	15 years 3 months	Bilateral medial hamstring lengthening, bilateral patellar tendon shortening, bilateral tibialis posterior lengthening	12 months	Bilateral calcaneocuboid fusion (2 years 5 months), new problem	5 years
12 Male	II	12 years 5 months	Bilateral medial hamstring lengthening, bilateral distal femoral derotation osteotomy, bilateral tendoAchilles lengthening, right Baumann procedure, bilateral open adductor lengthening	3 years	No additional surgery	6 years
13 Male	I	18 years 1 month	Bilateral medial hamstring lengthening, bilateral tibialis posterior lengthening, left distal femoral derotation osteotomy	12 months	Left tibialis anterior tendon shortening (1 year 2 months), new problem	None
14 Male	III	13 years 2 months	Bilateral medial hamstring lengthening, bilateral patellar tendon shortening, bilateral tibialis posterior lengthening, left distal femoral derotation osteotomy	12 months	Bilateral open adductor lengthening (4 years 1 month), new problem	5 years

untreated patients and patients treated by nonoperative methods such as orthotics and botulinum toxin injections. Third, we had incomplete follow-ups for some patients because one patient missed the first postoperative gait analysis and two patients did not yet have the final examination. Fourth, our findings are likely relevant only to a specific group of patients with spastic diplegia for which we established specific surgical indications. Finally, we had no measure of function. However, we note most single-event multilevel surgery outcome studies have a retrospective study design and the evidence base is limited by only one published randomized clinical trial at the moment [1, 11, 12, 30, 33, 37, 42, 43].

We found severe gait dysfunction in patients with spastic diplegia can be improved for the short-term in one operative session by single-event multilevel surgery. These findings are in accordance with reported findings [10, 11, 40, 42]. In our study the preoperative gait profile score of 20° was 5.1° higher than reported by Thomason et al. [43]. References to the kinematic graphs and surgical prescription show the procedures for correction of excessive knee flexion and equinus contributed most to the overall surgical outcomes (Fig. 1). Changes in other movement analysis profile domains were made toward improved gait except for pelvic tilt, which increased only slightly (Fig. 2).

Increased anterior tilt was a negative outcome of one study of single-event multilevel surgery, and more work is required to further reduce this problem [18]. It also may be that study was underpowered and could not detect note-worthy changes in some of the movement analysis profile domains. The mean age (13.6 years) of the patients at the time of the single-event multilevel surgery in our cohort was older than patients in other studies (mean, 6.4 [12],

12.0 [16], and 8.5 years [26]). The mean interval of our study (0.8 years) between the first gait analysis and surgery was comparable to the interval reported by Gough et al. (0.7 years) [12]. The surgical complication rate of 1.1% in our series was lower than that reported by Rodda et al. (10 surgical complications in four of 10 patients) [28].

The need for additional procedures after single-event multilevel surgery for treatment of spastic diplegia was first reported by Aiona and Sussman [1, 41]. After short-term followup in our gait laboratory, nine patients had additional surgeries after single-event multilevel surgery for further gait corrections (Table 5), combined with plate removal (when indicated). In our study, additional surgery was indicated for 2/3 of the patients because of relapses (n = 4) or new problems (n = 5). This number of additional surgeries is similar to that of Thomason et al. [43], who reported 22 subsequent surgeries were needed in 12 patients. A reduction in gait profile score from 20.7° to 11.1° at mid-term followup indicated 47% improvement toward normal gait; major improvements in the movement analysis profile for hip flexion, knee flexion, ankle dorsiflexion, and foot progression contributed to this. It probably is unrealistic to expect a single episode of surgery to provide lasting correction of all musculoskeletal problems and enhance and preserve gait function in all patients with spastic diplegia. It also may not be appropriate to focus on a single-event paradigm in all subjects. Staging surgery (and offering revision surgery when necessary) in this study achieved 47% improvement toward normal gait, but to preserve the early improvements 2/3 of our patients required additional surgery. In our study, the gait profile score was used to measure the overall gait disorder. The median value of the gait profile score for healthy patients is 5.2°. The improvement in our patients represents reductions of 47% (from preoperative to short-term), 62% (from preoperative to mid-term), and 31% (from short-term to mid-term) in the difference between abnormal and healthy gait patterns. The gait profile score [3] can be calculated independently of the feature analysis on which the gait deviation index is based [3, 39]. The movement analysis profile summarizes much of the complex information contained in the kinematic data and shows which parameters contributed to the change of gait. Measuring the outcomes of surgery in patients with cerebral palsy is difficult. Other factors such as the motivation of each patient and their abilities to participate in the postoperative rehabilitation program are likely to be important, although these are more difficult to quantify.

Single-event multilevel surgery, including lengthening of the contracted agonist, antagonist shortening [29], and selected bony procedures with stable fixation [30–32], improves gait dysfunction in patients with spastic diplegia. To preserve early improvements however, 2/3 of our patients required additional surgery because of relapses or new problems. It is unknown whether the improvements will last for decades.

Acknowledgments

We thank Mary Sheedy and H. Kerr Graham, MD, FRCS (Ed), FRACS for help with preparation of this manuscript. In addition we thank Alejandra Speiser for help in copyediting the final version of this manuscript.

Appendix 1. The Basle Concept for Single-event Multilevel Surgery in Cerebral Palsy

Demographics and surgical prescription for each patient

1. Based on instrumented three-dimensional gait analysis
2. Preoperative botulinum toxin test injections in all muscles planned for lengthening
3. Single-event multilevel surgery: Bilateral simultaneous surgical correction of all bony and soft tissue deformities

Individual procedures

Calcaneal lengthening osteotomy (Evans) or calcaneocuboid fusion with lengthening were performed in patients with clinical and radiologic evidence of a midfoot break, depending on stability of the Chopart joint.

Fixed equinus deformities (involving gastrocnemius and soleus) were managed by open Z-lengthening of the Achilles tendon.

In patients with excessively long triceps surae, the Achilles tendon was shortened by open plication.

In patients with clinical and kinematic evidence of deficient tibialis anterior function, the tendon was shortened by fixing the tendon at the ankle level and pulling it down to the insertion.

In patients with spastic equinovarus deformities attributable to documented tibialis posterior over-activity, the tibialis posterior was lengthened by open Zlengthening below the medial malleolus.

When the fixed flexion deformity at the knee was less than 25°, distal medial hamstring lengthening and supracondylar extension osteotomy were performed.

When the fixed flexion deformity at the knee exceeded 25°, distal medial hamstring lengthening was performed 3 months before single-event multilevel surgery to avoid late recurvatum in gait.

When the fixed flexion deformity at the knee exceeded 40° and/or the knee extension lag was greater than 10°, patellar tendon shortening was performed.

When there was radiologic evidence of hip subluxation (a migration percentage greater than 30%) with coexisting acetabular dysplasia, reconstructive hip surgery was performed, including femoral varus derotation osteotomy and a modification of the peri-ilial pelvic osteotomy [27]. This was done before single-event multilevel surgery to avoid an overly long surgical session.

Fixed flexion deformity at the hip was considered secondary to walking in crouch gait and knee flexion and was not corrected primarily but allowed to resolve gradually during the rehabilitation period.

4. Rehabilitation

Reconstructive surgery at the foot and ankle level was managed by below knee cast immobilization for 6 weeks.

Acute shortening of the knee extensor mechanism was treated with 6 weeks immobilization in extension in a knee brace, limiting flexion to less than 40°.

Rotational osteotomies of the femur and tibia were fixed with LCP plates, allowing for early mobilization and weightbearing.

At 6 weeks postoperative, a 4- to 6-week period of inpatient rehabilitation was provided, including gait training, mobilization, and strengthening, or an intensive rehabilitation program with at least three sessions of physiotherapy every week was provided.

5. Short-term results (1 to 3 years followup)

Before removal of fixation plates, another three-dimensional gait analysis was performed to judge the effectiveness of the original intervention and to detect unresolved gait issues or new problems.

5. Second operative intervention

Removal of fixation plates and any additonal procedures as required

7. Mid-term result (4–6 years after surgery)

A full biomechanical assessment, including instrumented three-dimensional gait analysis.

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8

Chapter 9

Explaining the variability in improvements in gait quality as a result of single event multi-level surgery in cerebral palsy

Erich Rutz, Susan Donath, Oren Tirosh, H. Kerr Graham and Richard Baker

9

Abstract

Purpose: This is a study of all children with spastic diplegic cerebral palsy (Gross Motor Classification System levels II and III) who had single event multi-level surgery (SEMLS) at a single tertiary referral hospital between 1995 and 2008 to identify factors predicting improvement in gait quality as quantified by the gait profile score (GPS). 9 factors (5 dichotomous and 4 continuous, including preoperative GPS) that might be expected to predict outcomes were identified and univariate and multivariable analysis used to explore how these affected outcomes.

Scope: Data from 121 children were included. The mean improvement in GPS of 4.3° was 2.7 times the minimal clinically important difference. Univariate analysis suggested that preoperative GPS is a very strong predictor of improvement in GPS ($p < 10^{-5}$) and when this is considered as a covariate only GMFCS level ($p = 10^{-5}$) and having had previous surgery ($p = 0.026$) were found to be statistically significant predictors of GPS improvement ($p < 0.05$). Children of GMFCS level II improved on average by 2° more than those of level III once differences in preoperative GPS had been accounted for.

Conclusion: Children with the most abnormal gait patterns preoperatively, and hence those with the most potential to improve are those that improve the most and surgery has clearly been beneficial. Over a quarter of children show changes in GPS which were less than the MCID. The majority of these were those with the least abnormal gait patterns preoperatively and further research is required to establish whether and how such children benefit from SEMLS.

Introduction

Single event multi-level surgery (SEMLS) is becoming a more and more widely accepted technique for correcting orthopaedic impairments of children with cerebral palsy [1–4]. A recent systematic review concluded that there is now robust evidence for substantial improvements in gait dysfunction following SEMLS with much weaker evidence for improvements in gross motor function, gait efficiency and health related quality of life [5]. A number of studies [6–12] have reported improvements in gait quality in terms of gait indices such as the Gillette Gait Index [13], Gait Deviation Index [14] or Gait Profile Score [15–17]. These all reported statistically significant improvements. All show considerable heterogeneity within the cohorts at baseline and follow-up.

Two studies [6,11] contain sufficient detail to demonstrate that there is also considerable heterogeneity in how gait quality is affected by SEMLS across the groups. The purpose of this study was to thus investigate the outcomes of SEMLS for children with cerebral palsy from a single centre using the Gait Profile Score (GPS) and to identify the factors which might be predictive of outcome.

Methods

This was single centre, retrospective cohort study, investigating the outcome of SEMLS, followed by postoperative physiotherapy, in which the child's outcome was compared to their preoperative status using the Gait Profile Score (GPS) and Movement Analysis Profile (MAP)[16]. Ethical approval for this study was given by the Ethics in Human Research Committee of the institution No 23144C. The records of all children who had had surgery for gait correction at the Royal Children's Hospital between 1995 and 2008 were searched to identify children satisfying the following criteria:

- A diagnosis of bilateral spastic cerebral palsy [18]
- GMFCS levels II and III [19]
- Gait analysis before and more than 12 months after SEMLS.

Before any data was examined, the authors determined a list of parameters which they considered might be predictors of outcome. Dichotomous variables were GMFCS (II or III), private health insurance (yes/no, as a surrogate for socio-economic status as clinical pathways are similar), adverse events and prior gait correction surgery (both classified as yes or no). Continuous variables were age at surgery, number of surgical procedures and preoperative GPS. During the analysis two other variables were added; gender [following, 20] and date of surgery (months since the first child was operated upon) as it was expected that improving surgical and rehabilitation techniques might lead to improved outcomes.

2.1. Indications

The indication for SEMLS was principally to improve gait dysfunction. However, eight children had significant hip displacement (MP > 40%). The primary indication for surgery in these children was to stabilise the hips, with improvements in gait being a secondary goal. Eleven children had been assessed for possible selective dorsal rhizotomy (SDR) and were rejected by the rigorous screening process required for this procedure in our institution. As such these children were less than ideal candidates for SEMLS. The gait dysfunction in this group was in part related to spasticity and in part to fixed contractures or other musculoskeletal pathology. Selection of surgical procedures had been based on the diagnostic matrix, in which information from the patient's clinical history, physical examination, instrumented gait analysis, radiological examination and examination under anaesthesia are synthesised [21]. The technical details of the operative procedures have been described in previous publications and included a strong commitment to selecting the correct ''surgical dose'' [4,12,22,23].

Rehabilitation was provided by community physiotherapists. Children were discharged after a mean of five days in the hospital. Inpatient rehabilitation was not available.

Quantitative three-dimensional gait data had been collected using one of a number of Vicon systems but all with same marker placement protocol and post-processing through Plug-in Gait (Vicon, Oxford, United Kingdom). The GPS provides a single index of gait quality as the RMS difference between data from one gait cycle and averaged data from a control cohort of typically developing children taken across the 9 most clinically relevant kinematic variables [16]. The equivalent RMS differences for the individual kinematic variables are known as Gait Variable Scores (GVS) and can be displayed in a bar chart called the Movement Analysis Profile (MAP). GPS and MAP were calculated for both legs on four individual gait cycles. The median GPS and MAP were calculated for each leg and averaged to provide a single index for each child using GaitaBase, a web-interfaced repository for gait analysis data [24].

Linear regression analysis was used to determine whether each of the proposed predictors of change had an effect on the GPS improvement following surgery. The original analysis plan was to perform separate bivariate regression analyses for each predictor and select those found to be significant at $p < 0.05$ for a subsequent multivariable regression analysis. Preoperative GPS was found to be such a strong predictor ($p < 10^{-5}$), however, that the separate regression analyses were repeated for the other potential predictors with preoperative GPS as a covariate. Any predictors that showed a statistical significance of less than 0.05 for this regression were then used as inputs to the multivariable analysis. One of the advantages of the GPS is that it can be decomposed into individual gait variables scores (GVS) that constitute the MAP [16]. To account for variation in preoperative GPS the participants were grouped by preoperative GPS into three equal groups. Within these groups the median improvement in the different gait variables were calculated.

Results

One hundred and twenty one children with spastic diplegia, 48 girls and 73 boys of mean age 10.7 (standard deviation 2.7 years) were identified as fulfilling the inclusion criteria. All had had a preoperative gait analysis a mean of 7.3months before surgery (s.d. 6.0 months) and a subsequent postoperative analysis a mean of 1.3 after surgery (s.d. 1.0 year). GPS was 15.5° (s.d. 3.9°) preoperatively and 11.2° (s.d. 2.5°) postoperatively with a change of 4.38 (s.d. 3.7°). The change in GPS (ΔGPS) was statistically significant ($p < 0.001$). An MCID of 1.68 has recently been defined for the GPS [15]. On the basis of this 86 children (71%) improved (ΔGPS > 1.6°), 32 (26%) showed no evidence of clinically important rchange (1.6° > ΔGPS > -1.6°), and 3 (2%) had deteriorated (ΔGPS < -1.6°).

Results of the simple linear regression analysis and with pre-op GPS as a covariate are presented in Table 1. The two regression analyses give quite different results confirming the importance of considering Pre-op GPS as a covariate. Only GMFCS level and previous surgery showed statistically significant regressions when pre-op GPS is controlled for in this way. Table 2 represents the result of the multivariable regression involving these three predictors. The r^2 value for this is 0.69 and the RMS residual 2.1°. It can be seen that the effect of previous surgery is no longer significant reflecting a correlation between previous surgery and GMFCS level. Fig. 1 plots GPS improvement against pre-op GPS for GMFCS II and III separately.

Given the strength of this relationship children were divided into three equal groups on the basis of preoperative GPS to study the GVS comprising the MAP. None of the children could be described as mildly affected so children with the lowest preoperative

GPS (<14°) were labelled "moderate", the next third (14° < GPS < 18°) as "severe" and the most affected third (GPS > 18°) as "very severe". The median changes for each GVS (taken across both legs for all children) and quartile range are plotted in Fig. 2.

Table 1. Results of bivariate linear regression and linear regression with preoperative GPS as a covariate for the different potential predictor variables. Figures in bold are statistically significant ($p < 0.05$).

Dichotomous variables		**Proportions**	**Bivariate analysis**		**With pre-op GPS as covariate**	
			Effect (standard error)	***p*-Value**	**Effect (standard error)**	***p*-Value**
GMFCS level	II:III	66:34	-0.34 (0.72)8	0.641	2.11 (0.44)8	4.10^{S6}
Gender	Female:male	74:26	-0.75 (-0.75)8	0.285	-0.46 (0.43)8	0.297
Health insurance	No:yes	35:65	0.73 (0.71)8	0.306	0.32 (0.45)8	0.471
Adverse events	No:yes	10:90	3.17 (1.11)8	0.005	1.15 (0.72)8	0.112
Previous surgery	No:yes	19:81	1.01 (0.87)8	0.306	1.20 (0.53)8	0.026
Continuous predictors	**Mean**	**Standard deviation**	**Bivariate analysis**		**With pre-op GPS as covariate**	
			Effect (standard error)	***p*-Value**	**Effect (standard error)**	***p*-Value**
GPS pre-op (8)	15.5	3.8	-0.76 (0.06)8/8	$1\ 10^{S26}$		
Date of surgery (months)	72.0	39.0	0.00 (0.01)8/ month	0.670	-0.01 (0.01)8/month	0.092
Age at surgery (years)	10.7	2.7	0.33 (0.12)8/year	0.009	0.11 (0.08)8/year	0.188
Number of procedures	7.6	2.1	0.00 (0.16)8/proc	0.996	0.06 (0.10)8/proc	0.608

Table 2. Results of multivariable regression analysis of change in GPS on pre-op GPS and the two predictors that showing statistically significant ($p < 0.05$) correlations using separate linear regression analysis with pre-op GPS as a covariate.

Predictors	**Effect**	***p*-Value**
GMFCS level	2.00 (0.44)8	1×10^{-5}
Previous surgery	0.87 (0.50)8	0.081
GPS pre-op (8)	-0.85 (0.05)8/8	5×10^{-31}

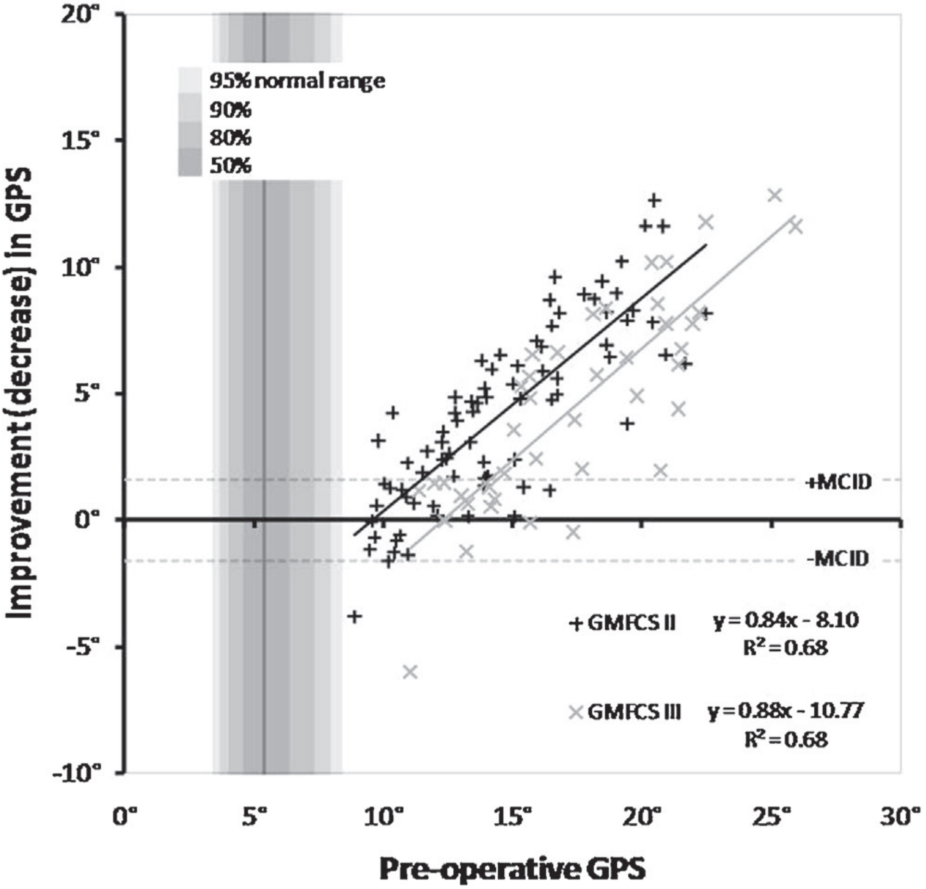

Fig. 1. Change in GPS plotted against preoperative GPS with children of GMFCS level II and III plotted separately. The horizontal dotted lines represent the minimal important clinical difference (MCID). Points plotted above the upper line represent children who can be said to have improved and points below the lower line are children who have deteriorated. Vertical bars represent ranges for children with no neuromusculoskeletal pathology.

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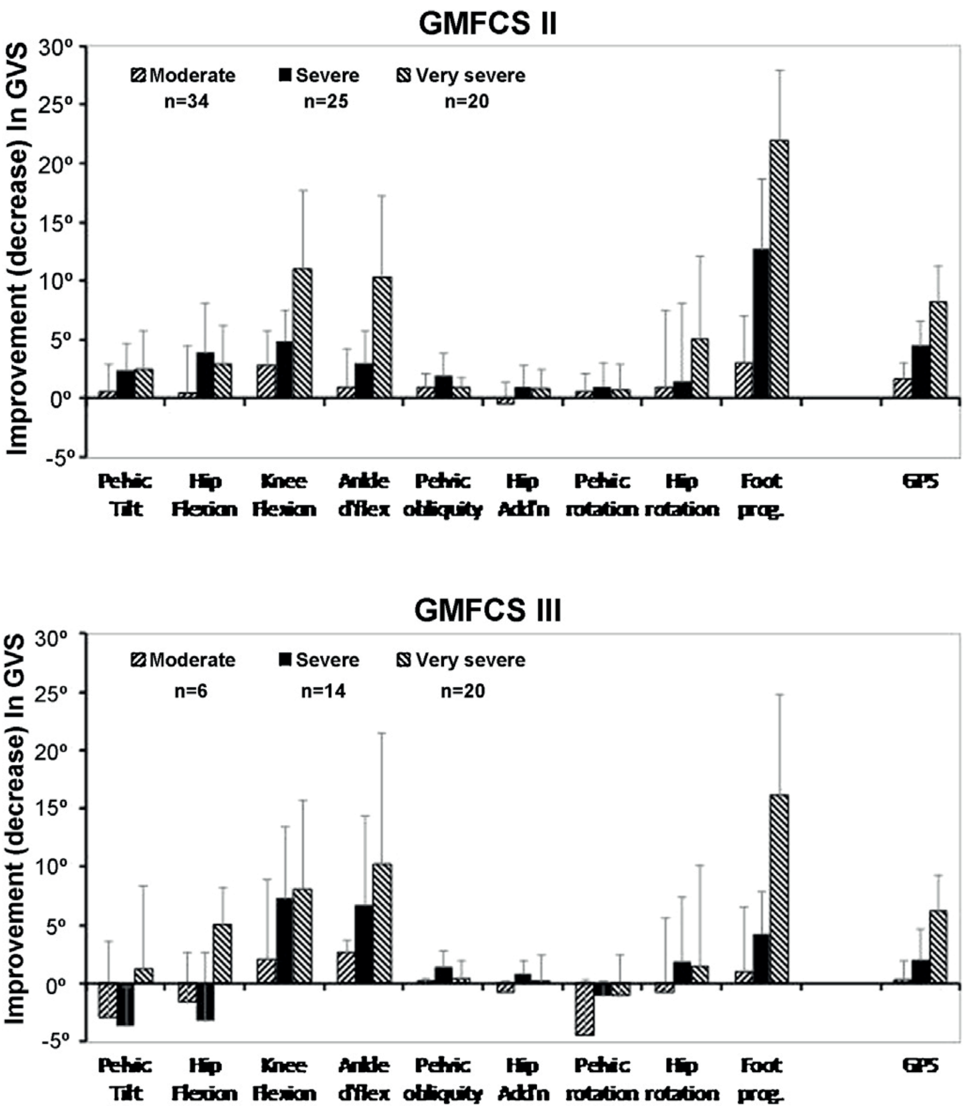

Fig. 2. Changes in the gait variable scores (GVS) comprising the movement analysis profile (MAP) stratified by preoperative GPS. ''Mod'' represents the third of all children with lowest GPS scores, ''severe'' those in the middle third, and ''very severe'' the third with the highest scores. Median and 3rd quartile range are plotted.

Discussion

The mean improvement (decrease) in GPS is 4.3°. This compares with the 4.6° decrease in GPS at 12 months reported from the randomised controlled trial of SEMLS conducted by Thomason et al. [12]. That study was conducted under near ideal conditions whereas this study represents all children who underwent SEMLS over a 13 year time period. This suggests that the excellent results reported from that trial are very similar to outcomes of routine clinical practice at the same hospital.

The regression analysis indicates that once pre-op GPS has been accounted for the only other predictor of improvement is GMFCS level. On average children at level II have a GPS 2° higher after surgery than those at level III with the same pre-op GPS. GMFCS is an indication of the severity of underlying neurology. It thus makes sense that the children at level II who are less affected have more potential for improvement as a result of SEMLS. The r^2 value suggests that 69% of variability in improvement can be attributed to GMFCS level and preoperative GPS and the low RMS residual (2.1°) confirms that once these factors have been accounted for the results of SEMLS are actually quite consistent.

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The relationship between improvement in GPS and pre-op GPS as illustrated in Fig. 1 is interesting. Improvement is clearly better in the children with more abnormal gait patterns preoperatively. This is unsurprising as these are the children who have the most potential for improvement. The data suggest that SEMLS will reduce GPS by about 86% of each degree above a threshold value (9.6° for GMFCS II and 12.2° for GMFCS III). These thresholds still fall well outside the range of GPS values for children with no neuromusculoskeletal pathology serving as a reminder that even after orthopaedic impairments have been removed that the children still have cerebral palsy and do not walk "normally".

Horizontal lines representing the minimal clinically important difference (MCID) for the GPS of 1.6° [15] are displayed in Fig. 1. 71% of children exceeded this and the average improvement was 2.7 times this value reinforcing the excellence of overall outcomes for the group. Correspondingly, however, 28% of children showed deterioration or improvement of less than this value and these are, predominantly, those who have the least abnormal gait patterns (lowest preoperative GPS) to start with. It is tempting to suggest that these children might be too good to improve from surgery but it should be noted that both values are considerably in excess of the 97.5th percentile of GPS scores for typically developing children (8.5°) and that all children were considered to have sufficiently poor gait patterns to warrant SEMLS which, by definition, involves at least four orthopaedic procedures. It is premature to extrapolate from this data to the conclusion that children with less abnormal gait patterns will not benefit from surgery. GPS is only one outcome measure. It has been shown to be mathematically related to the other recent indicator of gait quality, GDI, and highly correlated with GDI suggesting that it is unlikely that other measures of gait quality will show substantially different results. There is strong evidence, however, that without SEMLS children with CP are susceptible to some deterioration in gait quality [25– 27] and that surgery may act to prevent this. Factors other than gait quality may also be important, the most obvious of these being function which is not addressed by this study. For example some children with small changes in GPS, required less external support to ambulate after SEMLS, documented using the Functional Mobility Scale (FMS).

Fig. 2 shows that improvements in the GVS that constitute the MAP vary considerably with preoperative GPS. Again, children with the most abnormal gait patterns preoperatively are those who show most improvement. Improvement varies considerably across the different variables with foot progression, ankle dorsiflexion and knee flexion showing most evidence of improvement. For the third of children with the least abnormal preoperative gait patterns the variability in the GVS change is considerably greater than the mean improvement. This suggests that, in this group in particular, surgery leads to different but not necessarily more normal gait patterns. In line with the GPS analysis children in GMFCS II generally do better than those in GMFCS III. There is some evidence that within the moderate and severe groups SEMLS may be leading to deterioration in pelvic tilt, hip flexion and pelvic rotation for children in GMFCS III.

Understanding the relationship between preoperative GPS and change in GPS is important in understanding outcome studies. It is very clear from Fig. 1 that studies involving children with more abnormal gait patterns are likely to result in greater mean improvements than ones involving children with less involved gait patterns. Meaningful comparison of results from different studies is thus only possible if the range of preoperative GPS scores is clearly stated. In designing comparative studies stratification or minimisation approaches will be useful to ensure groups are matched for preoperative GPS and in analysing these it will be useful to include postoperative GPS as a covariate.

This is, of course, a factor affecting any clinical research in which a condition with considerable variation in severity is being studied and is not limited to use of the GPS.

Finally it is worth mentioning one statistical issue which is that the correlation between change scores and baseline data (as in Fig. 2) will be exaggerated if the measurement variability associated with the outcome measure is substantial. To investigate the likely effects of this a Monte Carlo analysis was performed which is fully reported in Appendix A. The findings of this where that measurement error is likely to lead to over-estimation the gradient of the regression line by around 4% and the intercept with the *x*-axis by less than a degree. Effects on r^2 and the *p*-value will actually be to reduce the strength of the correlation. Measurement error is thus very unlikely to influence either the presentation of the results or the clinical interpretation that has been placed upon it.

Assessing improvement after intervention amongst heterogenous groups of patients is very common in many fields of medicine. Investigating change scores as a function of baseline score has been demonstrated in this study to be lead to intuitively to an understanding of the underlying patterns in the data. This approach tends to be criticised as introducing a risk of misinterpretation of data as a consequence of the effects of measurement error. The Monte Carlo analysis described in Appendix A suggests that such effects have a minimal likely effect in this particular case and thus that such methods may be more useful than has been assumed. Further work will be required, however, to establish methods to determine the circumstance under which these risks are, or are not acceptable.

5. Conclusion

In summary we conclude that SEMLS can lead to substantial improvements in gait quality with a mean change in GPS of 4.3° (in comparison to an MCID of 1.6°) for children with cerebral palsy at GMFCS level II or III. The children with the most abnormal gait patterns show the largest improvements and surgery has clearly been beneficial. Over a quarter of children shows changes in GPS which were less than the MCID. The majority of these were those with the least abnormal gait patterns despite all of them still lying well outside normal ranges and being considered subjectively as having problems severe enough to require complex orthopaedic surgery. Further research is required to establish whether and how such children benefit from SEMLS.

Conflicts of interest

There are no conflicts of interest to declare.

Acknowledgements
The first author (ER) was financially supported by the Swiss National Science Foundation (SNF). The authors would like to acknowledge the contribution made to this research by the staff of the Hugh Williamson Gait Analysis Service by collecting the data on which the study is based. In addition we would like to thank Marianne Riksheim and Jo Røislien for statistical support.

Appendix A. Investigating potential effects of measurement error on the line of regression

Introduction

The results of the analysis reported in the full paper are that the outcome of SEMLS (change in GPS from baseline) correlates strongly with the preoperative GPS score. Some care is needed however in such an analysis if the measurement variability is substantial as this will lead to a correlation between change scores and baseline measures even if there is no treatment effect. To understand this consider that a given measurement of GPS is a consequence of some "*true*" score that is characteristic of the person and some *random* variability associated with the measurement process (this may include variability of the measurement itself and the performance of the individual on different occasions). Those people measured with high pre-operative scores are likely to be those with high true scores and for whom the random element was high as well. If they are re-assessed and the true score is the same but actual score is likely to reduce because the random element is unlikely to be so high on the second occasion. A similar effect means that low scores are likely to increase. The overall result is that there is likely to be a correlation between the change score and the baseline measurement even if the true scores are unchanged.

This can be illustrated by modelling the data. Preoperative GPS scores were represented by 41 points (the number of participants in the smaller GMFCS III group) uniformly distributed across the range of preoperatively recorded scores. No change was assumed and the postoperative scores were thus assumed to be equal to the preoperative scores. Measurement variability was then applied to both pre- and postoperative scores with a Gaussian distribution specified by its standard deviation. Fig. A1 shows the results of one such model in which variability with a standard deviation of 38 gives rise to an apparent correlation with R^2 = 0.39. Repeating this process 100 times gave an average slope of 0.29 and r^2 of 0.29. This suggests that the effects of this phenomenon are sufficiently large to merit further investigation.

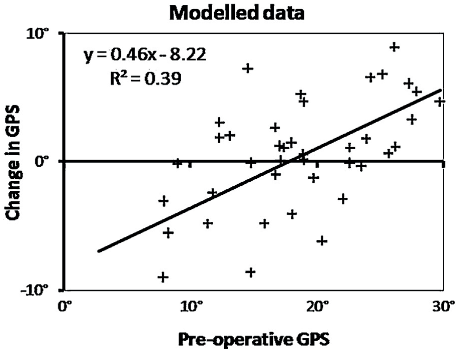

Fig. A1. Modelling of data with no true change between pre and postoperative scores but incorporating measurement error with a Gaussian distribution (SD = 38) to both measurements.

Method

A Monte Carlo analysis was performed to estimate the potential effect of measurement error on the data presented in this study. Clearly true scores are unknown so for the purposes of this analysis the measured values were taken as estimates of the true pre- and postoperative GPS scores and Gaussian error specified by its standard deviation was applied to both of these. The regression coefficient, p-value, slope and intercept with the *x*-axis where calculated for the relationship between the change score and the pre-operative score. This analysis was repeated one thousand times and the mean and standard deviation of the listed statistical parameters were calculated. The whole process was repeated for a range of error standard deviations between 0° and 4°.

Results

The results of the analysis are plotted in Fig. A2. When the variance is zero the analysis gives the same results as the original data as expected. As the variability rises there are increases in the slope and *x*-intercept as expected but these are very small. The effect of increased variability is to decrease r^2 value and increase the *p*-value suggesting that the potential for regression to the mean effects to reinforce the correlation is outweighed by the direct effect of increasing the variability about the line of regression.

Discussion

There have been no formal studies to estimate the variability of repeat measures of the GPS. Thomason et al. [12] reported a cohort of nine children fulfilling the eligibility criteria for this study who did not have study and were re-assessed after a 12 month period. The mean score for these children suggested a deterioration (average = 1.3°) over this period and the difference score thus may include some change in true score as well as measurement error. The RMS difference between pre- and postoperative scores for this cohort (2.3°) can therefore be taken as an upper bound for the measurement variability associated with the GPS over this time period. Reading from the graphs suggests that the likely effect of measurement error is likely to reduce the slope by less than 4% and increase the intercept by less than 1° from the values calculated directly from the measured results. These effects are very unlikely to influence the clinical interpretation that is based on the data. As commented above, the effect of measurement error is to reduce the strength of correlation as represented by r^2 and the *p*-value suggesting that the underlying correlation will be even stronger than reflected by r^2 and *p*-values calculated from the original data. The overall conclusion is thus that there is little potential for effects of measurement variability to significantly affect the findings of this study.

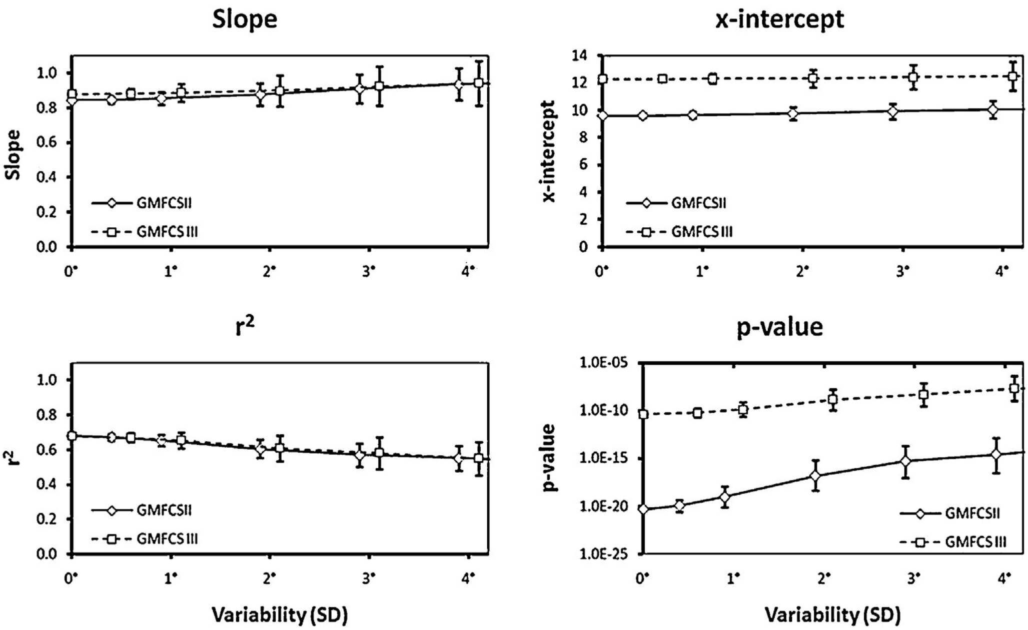

Fig. A2. Results of Monte Carlo analysis. Plotted value represent the mean from 1000 simulations and the error bars represent standard deviation.

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Chapter 10

General discussion

The aim of this thesis was to obtain insight in how different single-level or single-event multilevel surgery (SEMLS) procedures of the lower limb in children with unilateral and bilateral spastic Cerebral Palsy improve gait and to discuss the effects of general treatment principles of soft-tissue and bony surgery on gait and physical activity. For this purpose we have investigated results of surgical interventions at three levels i.e. the ankle joint, the knee joint, and the hip joint.

10.1. Summary of the results

In chapter 2, we described the results of weakening the muscle with botulinum toxin A (BTX-A) injections intended for surgery to improve walking. In our series of 110 consecutive patients with cerebral palsy (CP) considered for surgical muscle lengthening from 1999 to 2008, BTX-A was applied to identify patients at risk for functional deterioration. Gait analysis was repeated 6 weeks (maximum effect of BTX-A) and 12 weeks (follow-up) after the test injection to check for loss of joint control (excessive ankle dorsiflexion, knee flexion, or increased anterior pelvic tilt). After injections of the muscles intended for surgery, 20.9% (n = 23) showed deterioration in gait after preoperative BTX-A test injections. This percentage was in line with the percentage of deterioration in gait after surgery in the period before trial testing with BTX-A.

In chapter 3, we evaluated the results of combined Tendo Achilles lengthening and Tibialis Anterior Tendon shortening to treat equinus (plantar flexion position of the foot) in walking in 21 unilateral and 8 bilateral involved children with Cerebral Palsy, mean age 15.2 years. The gait scores improved for all patients, and 93% of the patients showed improved active ankle dorsiflexion in walking.

In chapter 4, the long term results of the surgical procedure of chapter 3 were described. Out of 29, 23 patients (mean age at index-surgery = 14.9 years) were included, there was a mean follow-up time of 5.8 years. 3 children (13%) have shown a relapse showing increased plantar flexion of the ankle in walking. The data of 12 children with spastic hemiplegia as well as 8 children with spastic diplegia has been analysed. There has been a significant ($p<0.05$) improvement in gait variable scores for ankle dorsiflexion (describes equinus and drop foot) of the operated legs versus not operated legs.

In chapter 5, the effect of surgical correction of gait on hip dysplasia has been evaluated retrospectively in all operated 11 children with unilateral CP. The gait parameters improved, but not the degree of hip dysplasia.

In chapter 6, the effect of surgical correction of fixed knee flexion contraction on hip flexion contraction has been evaluated retrospective in 12 patients with a follow up of 5 years after surgery. Hip extension improved in these patients without surgical intervention of the hip muscles.

In chapter 7, the stability of the Gross Motor Classification System (GMFCS) after surgery in 107 children with bilateral CP was evaluated in a retrospective cohort, classified as GMFCS level II or III. The mean age at surgery was 10 years 7 months (SD 2 y 8 mo). The primary outcome measure was the gait score. Changes in GMFCS level were studied at multiple time points before and after intervention. Gait dysfunction was partially corrected, with a mean improvement of 28% in the gait score. The GMFCS remained stable and unchanged in 95% of children and improved by one level in 5% of children.

In chapter 8, the effect of multi-level orthopaedic surgery was evaluated retrospectively of the data of 14 patients with spastic diplegia. At the time of surgery, one patient had gross motor function level I, 10 patients had Level II function, and three patients had Level III function, mean age of 13 years (range, 7-18 years). The minimum follow-up was 1 year (mean, 2 years; range, 1-3 years). At last follow-up, movement analysis profiles for knee flexion, for ankle dorsiflexion, and for foot progression improved as did the gait profile score. Additional surgery after the index procedure was performed in nine of the 14 patients because of relapse of the original or new gait problems. Major surgical adverse events occurred in one of the 14 patients.

In chapter 9, all children with spastic diplegic cerebral palsy (Gross Motor Classification System levels II and III) who had single event multi-level surgery (SEMLS) between 1995 and 2008 were evaluated to identify factors predicting improvement in gait quality as quantified by the gait profile score (GPS). The results show that children with a higher GPS improved more than children with a lower GPS, while children with a lower GPS will have less possibilities to improve.

In the general discussion the following subjects will be addressed:
1. Methodological issues of the studies
2. Consequences of soft tissue surgery on the level of muscle function
3. The effect of growth on the outcome of surgery
4. The outcome measures used in the studies
5. Future prospects: prevention versus treatment

10.2. Methodological issues

All studies performed were based on retrospective analysis of the data of the patients. This method has a risk of bias by patients lost for follow-up or selection of patients for evaluation. Disappointed patients mostly do not show up for evaluation and additional treatment.

Also, in retrospective studies the process of selection of children for a surgical procedure may be variable and varying in time.

Cerebral Palsy is an umbrella diagnosis for a very heterogeneous group of children. Children in Gross Motor Classification System (GMFCS) level I are only limited in sports activity like jumping and running, level II has limitations in walking distance but is able to walk without walking aids. Level III is only able to walk outside with a walking aid, level IV uses a wheelchair in moving around, and level V is unable to move around independently only in very special electric equipment. Besides the varying level of mobility, 50% of CP children have learning disability, which influences the level of daily activities(1). The number of children in the studies is very limited with respect to this heterogenicity.

Most studies about evaluation of surgical procedures suffer of low level of scientific evidence because these studies are often retrospective. As a consequence, the conclusions from these retrospective studies have to be interpreted with caution.

10.3. Consequences of soft tissue surgery on the level of muscle function

In Cerebral Palsy (CP) the primary impairment is disturbed muscle activation due to disfunction of the brain (2). The disturbed muscle activation can cause disbalance of forces around a joint during an activity, like walking, resulting in an abnormal pattern of movement. The disbalance can be caused by hyperactivity of the agonist of movement and / or weakness of the antagonist of movement. During the growth, these persistent disbalances in muscle activity will cause secondary deformations, like muscle shortening, reduced passive range of motion in a joint and finally fixed joint contractures, like knee flexion contraction (no full knee extension is possible anymore) and hip contractures or dislocations.

Orthopedic surgery addresses the secondary consequences of this disturbed muscle activation. The problem is to determine if the secondary deformation is primary caused by hyperactivity of the agonist or weakness of the antagonist. In chapter 2, the use of Botulinum toxin A intramuscular injections in muscles intended for surgery is described as a method to find out if hyperactivity of the agonist of abnormal posture is the pathological background or weakness of the antagonist. If weakness of the antagonist is the underlying problem, also weakening of the agonist of movement will cause deterioration, in this case of walking.

Surgery on a muscle will weaken the muscle itself. This has been described for the M. Gastrocnemius, with a high risk of development of crouch gait (walking with full foot contact, flexion of hip and knee), which is related with loss of walking ability in puberty or early adulthood (3-5). After hamstrings lengthening, also increased weakness has been demonstrated (6). Because of reduced selectivity of movement in a spastic paresis, measurement of muscle strength in a clinical setting is difficult (7). The differences in results of SEMLS (single event multi-level surgery) described in chapter 9 could be caused by differences in underlying muscle strength: CP children GMFCS level III are weaker than children GMFCS II (8).

10.4. The effect of growth on the outcome of surgery

The secondary deformations in children with CP develop in the course of time during growth. The age at surgery is important in relation to the years to grow: the younger age the surgery is performed, the higher the risk on relapse. In chapter 8, nine of the 14 children needed additional surgery in the course of time after the index surgery. The number of patients in this study was too small to analyse the factor age on the results of the surgery. Actually, final evaluation of surgical procedures to improve walking can be done in early adulthood, when the bodyweight as adult has been achieved.

10.5. The outcome measures used in the studies

In chronic health conditions, the ICF-CY (international classification of functioning, disability and health for children and youth) is used to describe the consequences of a disease (9). The results of treatment should be described also on the level of activities (meaningful activities in daily life) and participation (social activities).

The evaluation in these studies are mainly limited to the level of body function. Gait pattern itself is not related to the level of mobility (as defined in the ICF). Only the GMFCS classification gives some information about the level of mobility, but a classification is not sensitive for change. In chapter 7, one child improved in GMFCS level, and all others were stable after a mean follow up of 5 years. However, the age range at the moment of surgery was 6-17 years. In young children, natural development is an unknown factor. Around puberty and early adulthood, the risk of deterioration is present, so the large age range limits the interpretation of the results. A meta-analysis of SEMLS concluded that gait parameters improve, but not gross motor function (according ICF-CY mobility level) (10).

10.6. Future prospects

Prevention versus treatment

The Swedish CP-register exists since 1993, and can be considered as the longest prospective cohort study. They presented as the result of systematically control and treatment 0% hip dislocation in their population versus a prevalence of about 12%, with reduction of the surgical procedures to 1/3 in comparison with the period before the start of the register. This is a strong indication that hip dislocation is preventable.

However, Swedish CP register has well documented the frequency and contents of the systematic control, but not the contents of the non-surgical treatment (paramedic therapy, orthotic treatment, aids for positioning sitting and lying, spasticity management).

The surgical procedures are well described. The main message is that by systematic control a negative development (e.g. hip lateralisation) can be discovered, and early surgical treatment with ongoing non-surgical treatment offers the best results (11-14).

The results of soft-tissue surgery for hip lateralisation is also related to the degree of hip-lateralisation (15).

As obviously secondary orthopedic malformations are to a high degree preventable, it is important to develop protocols for non-surgical treatment in relation to the GMFCS level, and to extend the CP-register with registration of all non-surgical treatments and spasticity management. Because of the heterogeneity of the CP-population and the results on the long term (actually adulthood), randomized controlled trials are not suitable research models to answer the question which treatment is the most effective for which children.

The tendency to develop muscle shortening and reduction of passive range of motion (PROM) vary between children. Even after disappearance of spasticity by a Selective Dorsal Rhizotomy surgery, this can persist (16).Muscle growth and muscle properties are different in children with Cerebral Palsy (2,17,18). Identification of these risk factors could be important for the development of protocols for treatment.

Improvement of orthopedic surgery

In future, there will be always large numbers of children with Cerebral Palsy who has no access to preventive treatment. For these children, orthopedic surgery will be necessary as last resort treatment. So, better understanding of human biomechanics during gait, by the use of musculoskeletal modelling (19,21) is a tool to improve.

Multi-body simulations are powerful tools to examine the effects of pathologic gait such as crouched gait in individual patients. These simulations allow study the effects of the musculoskeletal geometry, the kinematics of the joints, and of muscle-tendon properties on forces and joint moments that the musculoskeletal system can produce.

Understanding the contribution of individual muscles on flexion/extension of the joints in healthy subjects can give insight into possible effects of muscle weakness on ambulation. Arnold et al. reported on this basis, that weak hip extensors, knee extensors, or ankle plantar flexors might contribute to crouch gait just as abnormal forces generated by the iliopsoas or the adductors muscle group (19). Furthermore the contribution of individual muscles to pathological gait as compared to healthy ambulation can be quantified for example with respect to their influence on angular accelerations and the acceleration of the center of mass. Individuals who walk in a crouch gait utilize similar muscles to support the center of mass (soleus, vasti, gastrocnemii, glutei and rectus femoris), but different muscles for forward propagation in the single limb stance phase (gluteus medius and hamstrings) (19,20,21,22).

Further simulations by Arnold et al. (19) revealed that in crouch gait a sustained activity of the quadriceps and the ankle plantar flexors in stance phase, as needed to support the body against gravity, gives rise to large opposing accelerations in forward and backward walking direction at the same time, hence simultaneously accelerating and retarding ambulation (23).

Besides the individual muscle contributions musculoskeletal simulations provide an insight into the joint loads experienced by patients walking in crouch gait. A joint reaction analysis of a musculoskeletal model of gait in different crouch positions revealed, that the compressive tibiofemoral force in severe crouch gait (minimum knee flexion greater than 50°) is doubled as opposed to the load in unimpaired ambulation (24).

Musculoskeletal modelling calculations can compute many biomechanical variables that cannot be easily measured in vivo or that cannot be measured at all, yet these biomechanical variables can be of great importance to improve the treatment of crouched gait in cerebral palsy patients (19). Future research in this field aims on quantifying the underlying mechanisms of impaired ambulation to extend the understanding of the causes of crouch gait by improving and deepening the existing methods and by addressing further quantities like the metabolic cost of a crouch gait pattern.

Such models of simulation are very complex. But we started 5 years ago at University children's hospital, UKBB to introduce such tools. Finally we were able to publish first results on our research.

Finally, randomised controlled trials can be used to find out if new developed procedures are more effective in comparison with the "*golden standard*". The outcome measures have to be not only on the level of body function, but also on the level of activity and participation with long term follow-up till adulthood.

The final question for all treatments for children with cerebral palsy should be: does our treatment improve their level of activities and participation in comparison with the natural course in time?

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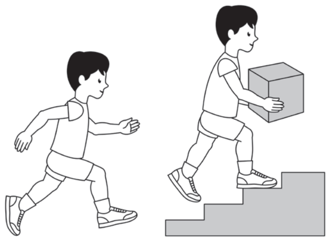
10

Appendix

GMFCS levels

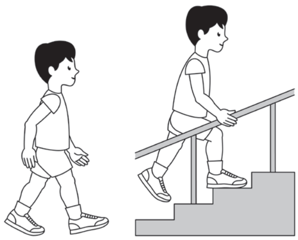
(with permission of Prof. HK Graham, Melbourne)

GMFCS E & R between 6th and 12th birthday: Descriptors and illustrations



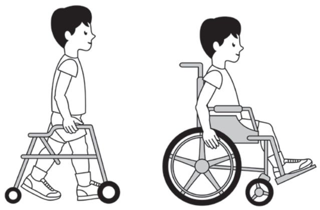
GMFCS Level I

Children walk at home, school, outdoors and in the community. They can climb stairs without the use of a railing. Children perform gross motor skills such as running and jumping, but speed, balance and coordination are limited



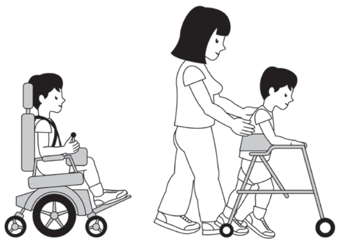
GMFCS Level II

Children walk in most settings and climb stairs holding onto a railing. They may experience difficulty walking long distances and balancing on uneven terrain, inclines, in crowded areas or confined spaces. Children may walk with physical assistance, a hand-held mobility device or used wheeled mobility over long distances. Children have only minimal ability to perform gross motor skills such as running and jumping.



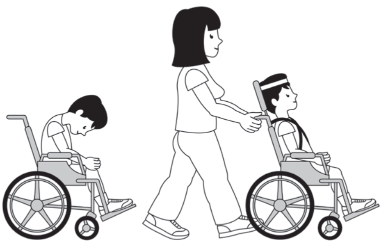
GMFCS Level III

Children walk using a hand-held mobility device in most indoor settings. They may climb stairs holding onto a railing with supervision or assistance. Children use wheeled mobility when traveling long distances and may self-propel for shorter distances.



GMFCS Level IV

Children use methods of mobility that require physical assistance or powered mobility in most settings. They may walk for short distances at home with physical assistance or use powered mobility or a body support walker when positioned. At school, outdoors and in the community children are transported in a manual wheelchair or use powered mobility.



GMFCS Level V

Children are transported in a manual wheelchair in all settings. Children are limited in their ability to maintain antigravity head and trunk postures and control leg and arm movements.

GMFCS descriptors: Palisano et al. (1997) Dev Med Child Neurol 39:214-23
CanChild: www.canchild.ca

Summary

12

Cerebral palsy (CP) is the most common cause of childhood-onset, lifelong physical disability, and of motor dysfunction affecting children with an estimated prevalence of 17 million people worldwide. Gait abnormailities associated with CP range from mild impairments such as walking on their toes to more severe gait dysfunction such as crouched and in-toed gait. These gait problems can lead to pain, deterioration in ambulant function, skeletal deformity and joint instability. Therefore the term „CP“ refers to a group of disorders of the development of movement and posture rather than to an aetiological diagnosis. There are many identified contributors to this disorder. However the cause remains unkown in about 15% of the cases.

Clinical management of children with CP is directed towards maximizing the function of the body, activities and participation and in contrast minimizing the effects of the factors that implement further deterioration of gait. Treatment strategies can consist of a variety of options such as: orthotics, physical therapy or orthopedic surgery. In an interdisciplinary team other options such as pharmaco-therapeutic treatment (e.g. Botulinum toxin type A or Intra-thecal Baclofen) and neurosurgical procedures (Selective Dorsal Rhizotomy, SDR, or Deep Brain stimulation) should be considered as well as treatment options.

For orthopedic surgical treatment there are many possibilities, but generally it includes corrections of bony deformities and soft tissue procedures. Often several surgical interventions are performed in one session, i.e. Single-event Multilevel Surgery approach (SEMLS). Therefore, SEMLS is defined as at least four surgical procedures, performed on two different anatomic levels (hip, knee, or foot and ankle) and on both sides of the body. The surgical procedure does not need to be symmetrical, but individually customised to the child’s needs. If the orthopedic problem is at one level, only, so called single level surgery might be sufficient as a less complex treatment option to fix the problem in one surgical session.

In chapter 2, we described the results of weakening the muscle intended for surgery to improve walking with botulinum toxin A (BTX-A) injections. In our series of 110 consecutive patients with CP considered for surgical muscle lengthening from 1999 to 2008, BTX-A was applied to identify patients at risk for functional deterioration. After injections of the muscles intended for muscle lengthening surgery, 20.9% (n = 23) showed deterioration in gait after preoperative BTX-A test injections. This percentage was in line with the percentage of deterioration in gait after surgery in the period before trial testing with BTX-A.

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In chapter 9, all children with spastic diplegic cerebral palsy (Gross Motor Classification System levels II and III) who had single event multi-level surgery (SEMLS) between 1995 and 2008 were evaluated to identify factors predicting improvement in gait quality as quantified by the gait profile score (GPS). The results show that children with a higher GPS improved more than children with a lower GPS, while children with a lower GPS will have fewer possibilities to improve.

In summary we were able to demonstrate that single event multi-level surgery or single level surgery is able to correct gait deviations in children with Cerebral Palsy. Finally, randomised controlled trials can be used to find out if new developed procedures are more effective in comparison with the "golden standard". The outcome measures have to be not only on the level of body function, but also on the level of activity and participation with long term follow-up till adulthood.

12

Acknowledgements

There are many people I would like to acknowledge for their patience, assistance and support throughout my doctoral candidature.

I extend gratitude to my dear wife Daniela, and to my two wonderful daughters, Sonja Helen Victoria & Julia Alana Patricia. I dedicate my thesis to my family who constantly reminds me how beautiful live is with them.

My parents, Brigitte & Hansjörg Rutz, and my brother Martin always supported me. Therefore a special thank is well deserved.

In addition I would like to thank my two supervisiors, Prof. Dr. Richard Jaspers, PhD, and Prof. Dr. Jules Becher MD, PhD.

A special thanks is deserved to the gait lab team in Melbourne, Australia, in particular to Prof. H Kerr Graham who was my mentor during the last ten years and to his former PA Mary Sheedy.

I would like to acknowledge the team in Basle, Switzerland, Prof. Dr. Reinald Brunner and the colleagues Dr. Bernie Speth, Dr. Thomas Schlemmer and PD Dr. Carlo Camathias.

In addition I wanted to thank to the orthotechnicians in Switzerland for the longstanding good and productive collaboration, in particular to my friend Res Reinhard from Ortho TEAM, Bern.

As of March 1st 2021 Dr. Erich Rutz has been supported by the Bob Dicken's Research Fellowship in Pediatric Orthopedics, University of Melbourne.

A very big thank goes to the children and their families who kindly participated in my research studies. I am truly indebted and grateful for their involvement.

List of publications

(corresponding to chapter numbers):

2: Preoperative botulinum toxin test injections before muscle lengthening in cerebral palsy

Preoperative botulinum toxin test injections before muscle lengthening in cerebral palsy.
Rutz E, Hofmann E, Brunner R. 2010 J Orthop Sci 15:647-53.

3: Tibialis anterior tendon shortening in combination with Achilles tendon lengthening in spastic equinus in cerebral palsy

Tibialis anterior tendon shortening in combination with Achilles tendon lengthening in spastic equinus in cerebral palsy.
Rutz E, Baker R, Tirosh O, Romkes J, Haase C, Brunner R. 2011 Gait Posture 33:152-7.

4: Long-term follow-up after tibialis anterior tendon shortening in combination with Achilles tendon lengthening in spastic equinus in cerebral palsy

Long-term follow-up after tibialis anterior tendon shortening in combination with Achilles tendon lengthening in spastic equinus in cerebral palsy.
Kläusler M, Speth BM, Brunner R, Tirosh O, Camathias C, Rutz E. 2017 Gait Posture 58:457-462.

5: Mulitlevel surgery improves gait in spastic hemiplegia but does not resolve hip dysplasia

Multilevel surgery improves gait in spastic hemiplegia but does not resolve hip dysplasia.
Rutz E, Passmore E, Baker R, Graham HK. 2012 Clin Orthop Relat Res 470:1294-302.

6: Hip flexion deformity improves without psoas-lengthening after surgical correction of fixed knee flexion deformity in spastic diplegia

Hip flexion deformity improves without psoas-lengthening after surgical correction of fixed knee flexion deformity in spastic diplegia.
Rutz E, Gaston MS, Tirosh O, Brunner R. 2012 Hip Int 22:379-86.

7: Stability of Gross Motor Function Classification System after single-event multilevel surgey in children with cerebral palsy

Stability of the Gross Motor Function Classification System after single-event multilevel surgery in children with cerebral palsy.
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8: Are results after single-event multilevel surgery in cerebral palsy durable?

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