MINI-SYMPOSIUM: CEREBRAL PALSY

(ii) The management of spastic diplegia

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Abstract  A consequence of improved survival of very premature babies is increasing numbers of children presenting to Orthopaedic surgeons with spastic diplegia. The majority of these children can walk but gait abnormalities are common and the natural progression is from muscle spasticity to contracture and eventual bony deformity. This results in increasingly inefficient gait as the child grows and gains weight. Intervention is initially by spasticity management, enhanced (over recent years) since the advent of Botulinum toxin type A, and later surgical intervention to both contracted muscles and bony deformities. Instrumented gait analysis is a prerequisite of surgery and all deformities should be addressed simultaneously. Surgery should only be considered once the child’s function has plateaued, usually between the age of seven and ten. Satisfactory results require patient and carer compliance in what is an intensive rehabilitation involving a multidisciplinary approach.

DEFINITIONS

Cerebral palsy (CP) is not a specific diagnosis but rather a collective term, used to describe a large group of children suffering from a variety of motor impairments, caused by non-progressive lesions in the immature brain. It has three key features:

- CP is a disorder of movement and posture,
- CP is caused by a non-progressive lesion in the immature brain,
- CP results in progressive musculoskeletal pathology.1

While the underlying brain lesion is static, the musculoskeletal manifestations are progressive. This definition satisfies orthopaedic surgeons as well as neurologists and paediatricians who must differentiate CP from various progressive disorders, such as the leucodystrophies. Although there are many classifications and subdivisions of the cerebral palsy syndromes, the classification of the movement disorder and the topographical distribution are the most useful.

CLASSIFICATION OF THE MOVEMENT DISORDER2

- Spastic 85%
- Dyskinetic (dystonic-athetoid) 7%

Spastic cerebral palsy is by far the most common type and is the principal movement disorder for which orthopaedic surgeons can provide useful help. With the improvements in the care of premature babies and the survival of very low birth weight neonates, the incidence of spastic motor disorders has increased.

TOPOGRAPHICAL CLASSIFICATION

- Monoplegia: involvement of one limb
- Hemiplegia: involvement of one side of the body
- Diplegia: involvement of both lower limbs with minimal involvement of the upper limbs. Spastic paraplegia implies no upper limb involvement.
- Triplegia: involvement of one side of the body, as in hemiplegia, combined with involvement of the contralateral lower limb. The lower limb involvement is always asymmetrical.
- Quadriplegia: involvement of all four limbs and the trunk. The alternative name is ‘whole body involvement’.

Spastic hemiplegia, spastic diplegia and spastic quadriplegia are the most common types. The vast majority of children with spastic diplegia have good cognitive abilities and will walk independently in the community.
A significant number remain dependent on assistive devices, walking sticks, crutches or a walker.

CNS PATHOLOGY

The classic brain lesion in spastic diplegia is periventricular leucomalacia, strongly associated with prematurity, often resulting in a relatively pure motor disorder with well-preserved cognitive abilities. Clinical examination usually reveals weakness, impaired motor control and spasticity in the lower limbs with deficits in fine motor skills in the upper limbs. Cerebral Palsy is the most common cause in childhood of the 'Upper Motor Neurone' (UMN) syndrome, which has a number of positive and negative features. The positive features are spasticity, hyper-reflexia and co-contraction. The negative features are weakness, loss of selective motor control and deficits in balance and co-ordination. The positive features are more amenable to intervention than the negative features. However, the prognosis for long-term function, especially walking, is much more closely related to the negative features than the positive features (Fig. 1). Strengthening is much more difficult to achieve than relief of spasticity and correction of deformities.

MUSCULOSKELETAL PATHOLOGY

Spastic cerebral palsy is usually described as 'short muscle disease' since spasticity and reduced voluntary activity result in impaired longitudinal growth of skeletal muscles. Throughout childhood there is a tendency for the growth of the muscle-tendon units to fall behind the growth of the neighbouring long bone which results in fixed contractures, secondary bony torsion and joint instability. The management of children with spastic cerebral palsy is therefore an effort to try and keep the growth of muscle tendon units more closely aligned to the growth of the neighbouring long bone. Two periods of intervention are required for these children. The first is a period of spasticity management, followed by surgery for contractures and bony torsional problems (Fig. 2).

SPASTICITY MANAGEMENT

Although the focus of this article is the orthopaedic management of children with spastic diplegia, orthopaedic surgeons must understand the principles of spasticity management and it is helpful if they become involved in the child’s management during this period. Children with spastic cerebral palsy have stiff muscles. Younger children have stiffness, which is usually reversible under anaesthesia and variable according to the child’s mood and many other factors. Spasticity can be assessed by measuring the ‘dynamic joint range’. This was introduced and referred to by Tardieu in the French literature and specific tests at the ankle, knee and hip were further described and developed by Boyd et al. Dynamic joint range of motion is measured by moving the joint rapidly through a passive range of motion, with the deliberate intention of activating the stretch reflex. The joint angle is recorded at that point, of first ‘catch’. The test is then repeated slowly to avoid activating the stretch reflex and the second joint range of motion is recorded. The first position is known as R1 and the second as R2. The greater the difference between R1 and R2, the greater the degree of spasticity, and the greater the need is for spasticity management rather than contracture surgery.

Options in spasticity management for children with cerebral palsy include oral medications, intramuscular injections of Botulinum toxin, injections of Phenol to motor nerves, selective dorsal rhizotomy and intrathecal Baclofen. The latter, administered by an implantable pump, is usually employed for children with spastic quadriplegia with very severe generalized spasticity.

SELECTIVE DORSAL RHIZOTOMY

Selective dorsal rhizotomy (SDR) is useful in a small subgroup of children with spastic diplegia who have moderate to severe spasticity, good underlying strength, good cognition, good family support and access to excellent physiotherapy and rehabilitation. The indications for SDR can be summarized as:

- Spastic — usually quite severe and generalized lower limb spasticity.
- Strong — good underlying muscle strength.
- Selective control — selective motor control should be present.
- Smart — good cognitive abilities to co-operate with demanding rehabilitation.
- Supported — good family and physiotherapy support are essential for success.

These criteria are predictive of a good outcome for any management programme, not just SDR. Selective dorsal rhizotomy has been widely used in a number of centres in South Africa and North America with excellent results being reported. In Europe and Australasia results have been more mixed and the procedure is much less popular. Significant complications may occur including progressive orthopaedic deformities, scoliosis, lordosis, hip subluxation and progression of foot deformities. In addition, the majority of children will still need multilevel orthopaedic surgery for contractures and bony torsional problems.
Intramuscular injections of Botulinum toxin A have been used for the past 10 years for children with spastic diplegia and there is good evidence in randomized clinical trials for a useful treatment effect. Injection of Botulinum toxin A results in a temporary chemo-denervation of muscle, improves muscle length and joint range of motion, improves tolerance to AFOs, delays the progress of contractures and will often enhance function. The principal indication for injections of Botulinum toxin A in spastic diplegia, is child who is toe-walking (dynamic equinus) aged between 2 and 6 years. There should be little or no contracture in the gastrocsoleus. Injection of Botulinum toxin A can be followed by serial casting, for mild fixed contractures, with good effect. Injections to other muscles including the hamstrings, adductors and iliopsoas are sometimes indicated, with some centres adopting a multilevel injection approach and reporting

**Figure 1** Although spastic diplegia is the result of a brain lesion, typically periventricular leukomalacia (PVL), a combination of disinhibition of the lower motor neuron results in the positive features of the upper motor neuron syndrome such as spasticity. However, it is the loss of connections to the lower motor neuron which leads to the negative features of the upper motor neuron syndrome of which weakness is the principal manifestation. The interaction between spasticity and weakness leads to both neural and mechanical changes in muscle and progressive musculoskeletal pathology. (Reproduced by permission of the Journal of Bone and Joint Surgery.)
There is growing evidence that the use of Botulinum toxin A allows orthopaedic surgery to be deferred until a later age and more appropriate disease stage. The need for isolated calf surgery can be completely abolished by the use of Botulinum toxin in early childhood thus avoiding iatrogenic crouched gait, formerly a ubiquitous problem.6,8

GAIT PATTERNS IN SPASTIC DIPLEGIA

Children with spastic diplegia are usually referred to orthopaedic surgeons because of a gait disorder. Although clinical experience and time spent in the Gait Laboratory will confirm that there are an infinite number of patterns of gait disorder, it is worthwhile looking for common patterns. The muscle groups, which exhibit spasticity and then contracture, have their principal action in the sagittal plane and result in characteristic gait patterns. These muscles include the psoas, the hamstrings, the rectus femoris and the gastrocnemius. These are principally the ‘two joint muscles’. Muscles, which cross only one joint are much less likely to develop contracture than those that cross two joints. Spasticity and contractures in these two joint muscles, result in characteristic gait patterns, which we have previously classified as:

Group I: true equinus—equinus ankle, extended knee, extended hip.

Group II: jump gait—equinus ankle, flexed knee, flexed hip.

Group III: apparent equinus—plantigrade ankle, flexed knee, flexed hip.

Group IV: crouched gait—calcaneus ankle, flexed knee, flexed hip. (Fig. 3)9

Younger children with spastic diplegia usually walk on their toes in equinus with extended hips and knees. As they get older, and sometimes because of isolated surgery to the tendo Achiillis, they may begin to develop increasingly flexed postures. It is very important for orthopaedic surgeons to understand that the natural history of gait disorder in diplegia is for deterioration. The principal changes with age include a decrease in walking speed, increased flexion and increased stiffness.

Jump gait is probably the commonest sagittal plane gait deformity presenting to the orthopaedic surgeon. However it is important to recognize apparent equinus, where the children are walking on their toes, because of flexion contractures at the hip and knee, not because of a contracture of the gastrocnemius. The most common reason for crouch gait is isolated lengthening of the tendo Achiillis although excessive use of injections of Botulinum toxin A to the calf muscle, may also be causative. Crouch gait is the most difficult sagittal disorder to correct because these children are older, stiffer, and have greater disability. It is always more difficult to shorten a muscle that is too long (the gastrocnemius) than to lengthen muscles that are contracted, in this case the hip flexors and hamstrings.

Although there may be some gait deviations in the coronal plane, the principal deviations in spastic
Although every child with spastic diplegia has their own individual gait pattern, we have identified four simple sagittal plane patterns which are frequently seen. These gait patterns are determined by which muscles are dominant and have developed spasticity or contracture. They provide a template for planing surgical correction and orthotic management. (Reproduced by permission of the Journal of Bone and Joint Surgery.)

Figure 3

Common gait patterns: Spastic Dipleiga

(A,B) This patient is in crouch although there are only mild lower limb contractures. The reason for crouch gait is severe lever arm dysfunction. This patient has a combination of medial femoral torsion, lateral tibial torsion, midfoot collapse and severe symptomatic hallux valgus. Unless these bony lever arms are surgically corrected, crouch gait is likely to persist and deteriorate with time. However correction requires a large amount of surgery and it is better to prevent these severe deformities by surgery earlier in childhood. The combination of medial femoral torsion and lateral tibial torsion results in the malalignment syndrome and this is particularly noted on the right side. The knee faces medially and yet the foot progression angle is still external.
diplegia are found in the sagittal plane as above, and in the transverse plane.9 The transverse plane problems are intoeing, usually the result of medial femoral torsion or ‘inset hips’. Additional problems include malalignment syndrome where there is both medial femoral torsion and lateral tibial torsion (Fig. 4). Finally transverse and coronal plane deformities of the foot and ankle may also be seen. Equinovarus is sometimes seen but is much less common than in spastic hemiplegia. Equinovarus is the result of spasticity/contracture in tibialis posterior and or tibialis anterior. Equinovalgus is acquired as the result of disordered biomechanics (equinus, stiff-knee gait) and is not caused by spastic/contracted peroneal muscles.10 The management of equinovarus is therefore tendon lengthening/transfer. The management of equinovalgus is based on bony stabilization surgery as well as correction of the disordered biomechanics.

THE ROLE OF INSTRUMENTED GAIT ANALYSIS11

Instrumented Gait Analysis includes:

- Standardized physical examination.
- Two-dimensional (sagittal and coronal) video recording of gait.
- Three-dimensional kinematics.
- Three-dimensional kinetics.
- Dynamic electromyography.
- Energy studies.

Instrumented gait analysis has been increasingly used to help our understanding of gait disorders in spastic diplegia, to plan intervention and to assess outcome.11 It is difficult to practice good quality surgical care for children with spastic diplegia, without access to a motion analysis laboratory. It is the authors’ view that single event multi-level surgery is such a critical intervention for these children that preoperative assessment in the gait laboratory is essential. Some surgeons in larger centres will have direct access to the gait laboratory and this is obviously the most desirable arrangement. However, others may need to refer their children to a regional centre and depend on the reports and recommendations made by the Gait Laboratory staff.

BIOMECHANICAL STUDIES RELEVANT TO ORTHOPAEDIC SURGERY IN SPASTIC DIPLEGIA

A number of anatomical and biomechanical studies in recent years have given insights into the biomechanics of normal and spastic muscle function, in a way which clinical studies alone could not. The term ‘muscle imbalance’ is widely and loosely used to explain the origin of musculoskeletal deformities in spastic diplegia and as a basis for corrective surgery. In a study of muscle excursion and cross-sectional area, it was demonstrated that the gastrocnemius and ankle dorsiflexors have such different physical characteristics that they cannot be considered to be ‘in balance’, in either normal subjects or in children with CP.12 The plantarflexors are six times as strong as the dorsiflexors. The concept of lengthening the triceps surae ‘to balance’ plantarflexor and dorsiflexor function however remains very durable, even though it is manifestly incorrect. The plantarflexors of the ankle must be balanced against the ground reaction force (GRF) not the dorsiflexors. Lifting the foot and ankle during swing phase (dorsiflexor function) does not require a large moment. Push-off in terminal stance (plantarflexor function) requires a large muscle moment. The concept of muscle balance should be redefined as a requirement for balance between the three anatomical levels, hip, knee and ankle, at not a single level. Children who walk in equinus, with the knee in extension or recurvatum have calf dominance over the knee flexors. Children who walk in crouch with calcaneus at the ankle and flexion at the knee and hip have dominance of the hip and knee flexors over the calf.

From modelling of muscle lengths in crouch gait, we have learned that many children who walk with flexed knee gait have hamstrings which are of normal length. It is the psoas which is shortened and requires lengthening, not the hamstrings. Most surgeons have been guilty of doing too much hamstring lengthening and not enough psoas lengthening.14

SURGICAL PROCEDURES IN SPASTIC DIPLEGIA

The correction of musculoskeletal deformities, in children with spastic diplegia, for the correction
of gait deviations, should usually be performed in one session. Mercer Rang caricatured the serial correction of deformities as the 'birthday syndrome' and his insights remain pertinent.\textsuperscript{15} If the surgeon starts at the ankle, the correction of equinus gait by lengthening of the tendo Achilles, is often followed by crouch gait. Correction of crouch gait by hamstring lengthening may be followed by hip flexion/anterior pelvic tilt. Correction of the hip flexion by lengthening of the psoas still leaves a stiff-knee gait, requiring transfer of the rectus femoris. It is not possible to balance the sagittal plane motors, unless all contractures are dealt with simultaneously. Piecemeal correction, one anatomic level at a time, tends to produce new deformities with each 'correction.' The child suffers multiple hospitalizations for surgery/casting/rehabilitation throughout childhood and spends many birthdays in hospital or in casts. There is now a general acceptance that the majority of procedures should be performed in one operative session, requiring only one admission to hospital and one rehabilitation. However, neither the surgeon, nor the family should underestimate the difficulties of single-event multilevel surgery (SEMLS). SEMLS requires military style overplanning, meticulous surgery, effective peri-operative management and excellent rehabilitation (Fig. 5). The principal components of a successful SEMLS programme are:

- careful planning, based on clinical, radiological and gait analysis assessments,
- education and preparation of the child and family,
- peri-operative care, including epidural analgesia and expert nursing care,
- carefully planned rehabilitation,
- appropriate orthotic prescription,
- close monitoring of functional recovery,
- follow-up gait analysis at 12–24 months after the index surgery,
- removal of fixation plates,
- follow-up until skeletal maturity, for new or recurrent deformities.

The surgical team should consist of two experienced surgeons and two experienced assistants. None of the surgical procedures are particularly complex but a single surgeon is unable to perform 8–16 consecutive procedures without fatigue and diminished performance. Expert anaesthesia and pain management is essential. Epidural analgesia is required to make SEMLS acceptable on social and humanitarian grounds. Post-operative nursing care must be vigilant. The use of epidural analgesia carries risks of masking the signs of compartment syndromes and decubitus ulceration. The surgery is but a series of mechanical steps which correct deformity. In the first 6–9 months after surgery, children are more dependent and less functional than they were prior to surgery. A child who walks into hospital with a typical diplegic gait pattern, leaves hospital in a wheelchair with straighter legs, but may be unable to walk independently for weeks or even months. After SEMLS, dependency increases, walking speed falls dramatically and the energy cost of walking rises.

\textbf{Figure 5}  (A, B) This child has severe diplegia and is dependent on elbow crutches. She has contractures of the psoas, hamstrings and gastrosoleus. This is the 'jump gait' pattern and is the most common sagittal plane alignment found in children with spastic diplegia. After single-event multilevel surgery which included lengthening psoas at the brim of the pelvis, medial hamstring lengthening, and Strayer distal gastrocnemius recession, a good extension posture has been achieved and AFOs can now be worn. (Reproduced by permission of the Journal of Bone and Joint Surgery)
dramatically. Only a carefully tailored and carefully monitored rehabilitation programme can ensure that the child will reach a higher level of function.

Weight-bearing may commence after a few days if there has been no bony surgery, after 1–2 weeks if there has been femoral osteotomy with stable internal fixation or after a maximum of 3 weeks, if there has been more extensive reconstructive surgery at the foot–ankle level. Casts are only required after foot and ankle surgery. Removable extension splints may be used at the knee level after hamstring-rectus surgery. The goal is to achieve full extension of the knee, combined with regaining full flexion, so that the transferred rectus femoris does not become scarred and adherent in its new site.

New ankle–foot orthoses must be prepared for immediate fitting after cast removal, usually 6 weeks after surgery. The initial post-operative brace is usually a Ground Reaction or Saltiel AFO (GRAFO). The orthotic prescription must be carefully monitored throughout the first year after surgery. A less supportive AFO, such as a hinged or posterior leaf spring, may be introduced when the sagittal plane balance has been restored and

Figure 6  The two joint muscles which require musculotendinous recession in single-event multilevel surgery are: psoas at the brim of the pelvis, fractional lengthening of the medial hamstrings and distal gastrocnemius recession. In addition the majority of children with spastic diplegia will have a stiff knee gait and benefit from transfer of the rectus femoris to the semitendinosus.
the plantar-flexion, knee-extension couple is competent. We monitor functional recovery and orthotic prescription by a gait laboratory visit every 3 months for the first year after surgery and yearly thereafter. These visits typically involve a careful clinical examination by the physiotherapist who conducted the preoperative gait analysis and a two-dimensional video recording of gait. A full, instrumented gait analysis is not required at every visit.

Muscles are the motors, which act on skeletal levers to produce movements. The muscle-tendon units are often contracted and the bony levers are often malaligned with the line of progression during walking (lever arm dysfunction). The frequently used procedures are therefore muscle-tendon lengthenings, tendon transfers, rotational osteotomies and bony stabilization procedures. These include:

**Soft-tissue surgery: tendon lengthening** *(Fig. 6)*
- Lengthening of the psoas ‘over the brim’ (POTB).
- Percutaneous lengthening of adductor longus.
- Medial hamstring lengthening (MHS).
- Lengthening of the gastrocnemius aponeurosis (Strayer).

**Soft-tissue surgery: tendon transfers:**
- Transfer of rectus femoris to the semitendinosus.
- Split transfer of tibialis anterior (SPLATT) for the varus foot.

**Bony surgery: rotational osteotomies** *(Fig. 7)*
- External rotation osteotomy of femur.
- Internal rotation osteotomy of tibia.

**Bony surgery: joint stabilization**
- Proximal femur varus derotation osteotomy (VDRO).
- Os calcis lengthening.
- Subtalar fusion.

**Occasional procedures**
- Pelvic osteotomy.
- Fusion 1st MTP.
- Epiphyseodesis.

**PRINCIPLES OF SURGICAL TREATMENT**
Surgical decision making in cerebral palsy may be a complex process, often requiring a combination of clinical examination, instrumented gait analysis and examination under anaesthetic. It is important to recognize that the gait observed in clinic is a combination of primary abnormalities, which require correction and secondary or coping responses such as vaulting or circumduction. Distinguishing between primary and secondary abnormalities is often difficult or impossible using clinical examination alone and it is in this instance that gait analysis is very useful.

Muscle lengthening should only be performed for fixed deformities (present under anaesthesia) and not for dynamic deformities. These are more safely managed by injections of Botulinum toxin A. Unfortunately
lengthened muscle may be weakened; surgeons should use this powerful technique with care. Performing fractional lengthening of broad aponeuroses and intramuscular lengthening of tendons may minimize the deleterious effects of lengthening surgery. In addition, power generation can be increased by improving the mechanical advantage of the muscle, i.e. improving the lever arm on which it works. This may require bony surgery such as osteotomy or joint stabilization.

In subsequent sections we will describe characteristic deformities at various levels of the lower limb, the analysis of such deformities and their surgical indications. It is useful to consider deformities in relationship to how they effect what have been described as ‘the five priorities of gait’ which are:

1. Stability in stance.
2. Foot clearance in swing.
3. Appropriate prepositioning of foot for initial contact.
4. Adequate step length.
5. Conservation of energy.

Energy expenditure during walking is impossible to quantify by clinical examination but is readily assessed by measuring oxygen consumption and carbon dioxide production during gait analysis, using a portable metabolic cart, such as the Cosmed K4. By determining these parameters pre- and post-operatively, the results of surgical intervention can be assessed objectively. There are three principal mechanisms by which energy is conserved during gait:

- Minimizing excursion of centre of gravity.
- Control of momentum. During the second half of stance the ground reaction force (GRF) is maintained in front of the knee by eccentric contraction of the soleus. This produces an extensor moment, which stabilizes the knee without active contraction of the quadriceps—the plantar flexion-knee extension couple.
- Active or passive energy transfer between segments. This is largely accomplished by using those muscles, which cross two joints. For example, the rectus femoris acts concentrically at the hip to augment flexion during pre- and initial swing whilst at the same time it is acting eccentrically at the knee controlling flexion. It is in fact absorbing energy at the knee and transferring it to the hip.

Each of these mechanisms can be observed to be abnormal in many patients with cerebral palsy and they can often be rectified to some extent by surgical intervention or orthotic prescription. It should be remembered that when the energy cost of walking crosses a certain threshold (and this will vary between individuals) patients will cease to ambulate and become reliant on wheelchair mobility.

**Dynamic Ankle Function:**

**Ankle Rockers**

When attempting to analyse the ankle and foot in the sagittal plane it is useful to divide stance phase into three different ‘rockers’. First rocker commences with heel strike and finishes when the foot is flat on the ground. It is largely controlled by eccentric contraction of tibialis anterior and is invariably absent in spastic diplegia, even when involvement is very mild. Second rocker commences at foot flat and terminates at heel raise. During this phase the ankle dorsiflexes under controlled eccentric contraction of soleus and to a lesser extent gastrocnemius. Spasticity and contracture of the gastrosoleus limit dorsiflexion during second rocker. Third rocker lasts from heel rise to toe off and is the phase of maximum power generation, accomplished by concentric contraction of gastrocnemius and soleus. This accounts for approximately 45% of the power generation required for walking. In swing phase progressive dorsiflexion should be seen, the result of phasic activity of the ankle dorsiflexors, effecting clearance and pre-positioning the foot for initial contact of the next gait cycle.

Observation of the gait of patients with ankle equinus will reveal a number of different patterns depending on the severity of involvement and level of involvement of other levels. Considering the priorities for normal gait it is seen that persistent equinus interferes with priorities two and three during swing and one and four during stance. Step length is reduced because the second and third ankle rockers are curtailed in the stance limb. The range of ankle dorsiflexion should be tested dynamically and statically. The relative contribution of the gastrocnemius and soleus can be ascertained by comparing the range with the knee flexed (soleus) and extended (soleus and gastrocnemius). This forms the basis of the Silfverskiold test and has important implications when considering surgical intervention (see below).

Although most abnormalities around the foot and ankle occur in the sagittal plane it is also important to identify problems in the coronal and transverse planes. Hindfoot varus and valgus should be noted and the flexibility and corrigibility ascertained. Valgus is primarily in the subtalar joint but concomitant ankle valgus is common in more severely involved children. Standardized weight-bearing X-rays of the foot and ankle mortise are required in all children. Hindfoot valgus is often associated with breaching of the midfoot, lateral subluxation of the navicular on the talus, abduction of the forefoot and an increasingly external foot progression angle. This reduces stance phase stability and the ground reaction force is also mal-directed out of the plane of progression, resulting in abnormal stresses on proximal bones and joints. Excessive lateral tibial torsion is frequently found with the valgus/abducted foot and careful clinical and radiological assessment is required to determine how
much of each deformity is present, because this will determine the surgical prescription. Accurate clinical assessment of tibial torsion is notoriously difficult and we prefer to use a combination of measurement of the thigh–foot–angle (TFA) and the bimalleolar axis of the ankle, measured with the patient prone and knees flexed.

**FOOT AND ANKLE: SOFT-TISSUE SURGERY**

The gastrocnemius is always more contracted than the soleus in spastic diplegia and selective lengthening of the gastrocnemius is all that is required, for the majority of children.\(^\text{11,17}\) Even when a contracture of the soleus is present, differential lengthening of the gastrocnemius and soleus is better and this requires surgery in the calf, not the tendo Achillis (Fig. 8). Only very severe and neglected equinus deformity requires lengthening of the tendo Achillis. The White slide technique, performed under direct vision is a much more controlled and satisfactory procedure than the Hoke triple hemisection technique, performed percutaneously.\(^\text{18}\)

The main complication of this surgery is gradual failure of the plantar-flexion-knee extension couple, leading to calcaneus gait, which is more disabling and difficult to treat than the original problem. Surgeons rarely ‘over-lengthen’ the heel cord, but isolated lengthening of the tendo Achillis will result in the gradual development of crouch gait in up to 40% of children with spastic diplegia.\(^\text{8}\) The ‘over-lengthening’ is mediated by biomechanical changes and growth, not surgical imprecision. When the GRF falls behind the knee, the soleus responds to the continual stretch by adding more sarcomeres, in series. In time, the soleus becomes functionally too long, and biomechanically incompetent and calcaneus-crouch progresses rapidly.

We have shown that deferring the surgery until age 8 years, reduces the risks of both recurrence and over-correction.\(^\text{8}\) Isolated calf lengthening is contraindicated if
proximal levels are also affected, because this is the most frequent cause of crouch. The more proximal operations on the gastrocsoleus are the most stable and safest in terms of avoiding calcaneus. From proximal to distal, these are:

- Strayer distal gastrocnemius aponeurosis lengthening (GAL).\(^{17}\)
- Strayer distal gastrocnemius aponeurosis lengthening, (GAL) combined with soleal fascial lengthening (SFL).
- Baker and Vulpius lengthening of the conjoined gastrocnemius fascia.\(^{19,20}\)
- Lengthening of the tendon Achilles. (White, Hoke).\(^{18}\)

Varus deformity is invariably secondary to spasticity of tibialis posterior and/or tibialis anterior. Varus is easily over-diagnosed in the presence of intoed gait, secondary to medial femoral torsion, a phenomenon described as ‘roll-over varus’. When in doubt, first correct the femoral torsion and later re-evaluate the foot and ankle. Intramuscular tenotomy of the tibialis posterior is usually all that is required for mild dynamic varus.\(^{21}\) A few children with more severe deformities require a split transfer of tibialis anterior to the lateral side of the foot. There is no role for either tenotomy of tibialis posterior or a complete (inter-osseous) transfer. Both procedures will result in severe valgus.

**SURGICAL TECHNIQUE (STRAYER)\(^{17}\)**

The patient is in the prone position for many of the component parts of multilevel surgery, including Strayer calf-lengthening. A posterior midline incision, 3–4 cm long, is made, centred over the musculotendinous junction of the gastrocnemius and its aponeurosis. The deep fascia is divided longitudinally and the sural nerve and short saphenous vein are identified between the two bellies of gastrocnemius and protected. A small longitudinal midline incision (approx 0.5 cm) is made in the gastrocnemius aponeurosis using sharp dissection. This is carefully deepened until the plane between the gastrocnemius aponeurosis and the soleus fascia is identified. The plane is then fully developed using a combination of finger and blunt dissection (Fig. 8). Once the two layers have been separated, the aponeurosis of gastrocnemius is divided transversely, and the muscle bellies are allowed to recess proximally and are then sutured in the appropriate position. This gives increased ankle dorsiflexion and correction of the equinus deformity. If the range of dorsiflexion is still limited to less than plantigrade, with the knee in extension, the fascia overlying the muscle belly of soleus can be gently striped transversely. After wound closure, a below knee cast is applied, with the ankle at neutral. This remains in place for 6 weeks and is then replaced by an ankle-foot orthosis. This surgery is inherently stable and immediate weight bearing is encouraged.

**FOOT AND ANKLE: BONY SURGERY**

The most common deformity in spastic diplegia is equinus, which over a variable period usually becomes associated with hindfoot valgus and breaching of the midfoot. This may be partially correctable using orthotics initially but invariably becomes fixed and no longer amenable to bracing or soft-tissue surgery. The most appropriate surgery in these instances is bony stabilization, either by lengthening of the lateral column of the foot (os calcis lengthening) or extra-articular fusion of the subtalar joint.\(^{22,23}\) Os calcis lengthening corrects subtalar joint evasion and midfoot breaching by elongating the lateral column of the foot, driving the heel into varus and raising the medial arch. This procedure has the added bonus of maintaining joint motion. The indication for os calcis lengthening is a flexible valgus deformity of the heel in association with an abductus deformity of the forefoot, in a patient who walks independently. Arthrodesis of the subtalar joint is a reliable means of correcting hindfoot valgus and with secondary correction of the midfoot. It is useful for more severe deformities, in patients who require assistive devices and long-term orthotic support. We prefer a modified Fulford technique, utilizing a cannulated screw passed through the talar neck, across the sinus tarsi into the calcaneum, combined with iliac crest autograft (Fig. 9). Both os calcis lengthening and subtalar fusion can be usefully combined for the correction of more severe deformities and both will have the effect of improving the lever arm for gastrocsoleus. Correcting hindfoot valgus inevitably increases tension in an already tight gastrocsoleus and calf lengthening is often required in conjunction with foot stabilization.

Hallux valgus is commonly associated with deformities in the hindfoot, midfoot and proximal gait deviations, such as stiff-knee gait, which causes toe scuffing. It is pointless to try to correct hallux valgus in spastic diplegia by the usual menu of surgical options, when there are so many biomechanical factors which contribute to recurrence. The most reliable procedure is fusion of the first metatarsophalangeal joint, either in conjunction with, or after correction of, the proximal deformities. We favour a cup-and-cone reamer technique with plate and screw fixation.

**SURGICAL TECHNIQUE: OS CALCIS LENGTHENING\(^{22}\)**

An Ollier incision or a longitudinal incision can be used to approach the lateral aspect of the os calcis. The aim of the surgery is to perform an osteotomy parallel to and approximately 1 cm proximal to the calcaneo-cuboid joint. The osteotomy should be between the middle and anterior facets of the subtalar joint (Fig. 10). The osteotomy site is carefully distracted and a trapezoid
of autologous or allograft corticocancellous bone can be inserted. Post-operatively, non-weight-bearing in a below knee cast is advised for 3–4 weeks, followed by 3–4 weeks of weight-bearing. At 6–8 weeks after surgery, graft incorporation is usually adequate to permit cast removal and fitting of an AFO.

**FOOT AND ANKLE: ROTATIONAL OSTEOTOMY—SUPRAMALLEOLAR OSTEOTOMY OF THE TIBIA**

Lateral tibial torsion may occur in isolation or in conjunction with medial femoral torsion. In isolation, lateral tibial torsion results in an external foot progression angle and 'lever arm dysfunction' because the foot lever is effectively shortened and mal-directed, in relation to the line of progression. However, when lateral tibial torsion and medial femoral torsion co-exist the foot progression angle may be relatively normal but the knees face inwards and there is still very significant lever arm dysfunction (malalignment syndrome). Derotational osteotomy of the tibia is an effective means of addressing this problem. We prefer to perform a very distal, supramalleolar osteotomy, which increases the cross-sectional area of contact between the two osteotomy surfaces. This increases stability, which facilitates early weight bearing, minimizes the risk of secondary deformities and is associated with reliable and rapid union. In order to gain the desired degree of rotation (which can be accurately determined using transverse plane kinematic data on gait analysis) the fibula should be divided at the same level as the tibia. We stabilize the osteotomy with AO/ASIF small fragment, contoured T-plates. Concomitant tibial osteotomy and bony foot surgery should be undertaken with caution. Severe post-operative swelling may occur and result in wound dehiscence.

**KNEE**

The knee functions primarily as a hinge joint and the principal gait deviations in spastic diplegia are found in
the sagittal plane. The vasti are active during the first 20% of stance phase, acting as shock absorbers and stabilizing the knee. After this point the calf muscles provide stability by restraining forward movement of the tibia, thereby maintaining the ground reaction force in front of the knee and activating the plantar-flexion knee-extension couple.

During stance phase, most abnormalities are secondary to spasticity or contracture of the hamstrings, which limit knee extension. An obvious consequence of this is difficulty with clearance of the contralateral swing phase limb. This may require a secondary compensation such as circumduction. If the knee does not extend fully, the ground reaction force during mid-stance will pass behind the knee and the plantar-flexion knee-extension couple will not be activated. Stability will depend on continued activity of the quadriceps, significantly increasing energy expenditure. In the long term this may lead to patella alta and patellofemoral pain, due to excessive loading. A further consequence of excessive knee flexion in stance, is the requirement for excessive hip flexion. The interrelationships between excessive hip and knee flexion are complex. If the hamstrings are contracted or the quadriceps relatively weak, excessive knee flexion may occur throughout stance resulting in crouch gait. If less severe, the patient may demonstrate ‘jump knee’ where there is excessive knee flexion at initial contact, followed by relatively good extension in late stance.

Another stance phase knee gait deviation is recurvatum, which is invariably the result of previous hamstring lengthening, in association with a spastic or contracted gastrocnemius. The plantar-flexion knee-extension couple is overactive.

During normal gait approximately 60° of knee flexion are required during swing phase. It is important that peak knee flexion occurs sufficiently early in swing phase, to allow clearance of the foot, which is still plantar-flexed at this point. In swing phase most problems relate to inadequate knee flexion. The rectus femoris is usually only active during late stance and initial swing but in cerebral palsy is often active throughout swing phase. The consequences of this are a ‘stiff knee’ gait due to co-contraction of the rectus femoris and medial hamstrings.11,25

Sagittal kinematics may enable identification of one of the four different abnormalities mentioned above. Measurement of the static range of knee motion is critical, particularly assessing any fixed flexion, a key feature of decompensation.

Hamstring spasticity and contracture are best evaluated by assessing the popliteal angle. The rectus femoris is examined using the Duncan Ely test, with the patient in the prone position.11

Knee: soft tissue surgery

Crouch gait and hamstring contracture are readily identified but the temptation to lengthen hamstrings in isolation should be resisted unless other levels, particularly

Figure 10  (A, B) Pes planovalgus is preferably corrected by os calcis lengthening. This procedure corrects the hindfoot valgus and forefoot abduction restoring the foot as a lever arm as well as a base of support for stability in stance.
the hip, are unaffected. If tight hip flexors are not addressed at the same time increased pelvic tilt and lumbar lordosis will result. It is easy to over-estimate the significance of hamstring contracture and easy to underestimate psoas contracture.

The medial hamstrings are more commonly and usually more severely contracted than the lateral. Fractional lengthening of both medial and, if required, lateral hamstrings can be accomplished through midline posterior incision just above the knee. The preferred technique includes intramuscular tenotomies of gracilis and semitendinosus and fractional lengthening of the semimembranosus, by performing one or two stripes through its broad aponeurosis. The semitendinosus may be harvested at the time of medial hamstring lengthening, for subsequent transfer of the rectus femoris.

When rectus is to be transferred, our choice is to detach it distally from the patella and transfer it medially to semitendinosus. In this position it has been shown to increase knee flexion during swing. Alternative recipients are gracilis, sartorius and the iliotibial band. It should be noted that early physiotherapy is imperative following this transfer, to stop adhesion formation.

Knee: bony surgery

If soft-tissue lengthening is insufficient to obtain complete knee extension bony surgery may be necessary. In skeletally immature patients anterior staple epiphysiodesis of the distal femur is an attractive alternative as it is associated with minimal morbidity and is immediately stable, allowing ambulation and physiotherapy to continue uninterrupted. Approximately 1° of correction per month can be expected until skeletal maturity. In older patients, an anterior closing wedge osteotomy of the distal femur may occasionally be indicated, combined with shortening of the patellar tendon. This is a major intervention, requiring a major rehabilitation programme.

Abnormalities of gait related to the hip may be considered in three groups: inadequate power, insufficient range of movement and malrotation. In spastic diplegia the hip flexors, adductors and internal rotators are stronger than the corresponding extensors, abductors and external rotators. In younger patients this may lead to dynamic deformity but over a period of time contrac-
tures develop. These are characteristically flexion and adduction. Lack of hip extension in mid-stance and terminal stance impedes forward progression, weight-bearing stability, and the advancement of the contra-lateral limb for adequate step length.

Patients with diplegia usually maintain better function in proximal muscles than distal. In normal gait, the majority of power for walking is generated by the gastrocnemius, during push off in terminal stance. In diplegia, patients rely more on proximal muscles especially the hip flexors, in effect using the hip to pull up. If these muscles are weak, gait may be profoundly affected. Weakness of the hip abductors will be manifest as a Trendelenburg gait, necessitating significant shifts of the trunk and centre of mass, and increasing energy expenditure. There will also be a reduction in stability of the stance phase limb and interference with clearance of the contralateral limb.

At birth, femoral neck anteversion measures 40° but this reduces to approximately 15° by skeletal maturity. Derotation results from pressure exerted on the femoral neck by Bigelow’s ligament, when the hip comes into full extension during normal gait. When the hip does not extend fully, derotation may not occur. The normal stimulus to proximal femoral remodelling is lost.

The typical gait of children with spastic diplegia is one of hip flexion, adduction and internal rotation. Foot progression may be internal or normal if lateral tibial torsion has developed. Transverse plane gait deviations are very difficult to recognize and quantify without instrumented gait analysis. For example, when the knees rub together it is usually the result of medial femoral torsion, not scissoring because of adductor spasticity. Femoral derotation osteotomy is required, not an aggressive adductor release. Pelvic retraction makes interpretation of knee position and foot progression angle very difficult. Examination of the hip joint should include assessment of flexion, abduction and the rotational profile. Thomas’s test can be used but the Staheli method is often more accurate in these patients. This requires the patient to be turned prone with the patients legs supported off the end of the examination couch. The most important assessment of hip rotation is with the hips extended. Again the patient should be prone with the knees flexed to 90°. The tibia then serves as a useful goniometer to measure internal and external rotation.
If hip subluxation is dealt with early, it is very unusual for both can be corrected by proximal femoral osteotomy. In this situation a rotational osteotomy of the femur permits the greater trochanter to remain in this advantageous position, in respect of the abductor lever arm, but the remainder of the lower limb is rotated back into the line of progression and the in-toed gait is corrected. When medial torsion and hip subluxation are present, both can be corrected by proximal femoral osteotomy. If hip subluxation is dealt with early, it is very unusual for children with diplegia to require pelvic osteotomy or open adductor releases. For the majority of children with diplegia who have normal hip development, in-toed gait and medial femoral torsion can be corrected equally well by a rotational osteotomy of the proximal or distal femur. The proximal osteotomy is usually performed with the patient in the prone position, when the rotational arcs of both hips can be easily checked before and during surgery. A 90° or 100° AO/ASIF blade plate is used to achieve stable fixation. We aim to correct anteversion to 0°–10°, leaving only 10°–20° of internal rotation at the hip. Distal osteotomy is performed with the patient in the supine position, a tourniquet can be used, surgery is faster and blood loss is lower. A subvastus approach is used to access the distal femur, a transverse osteotomy is performed and the corrected position held with an AO/ASIF dynamic compression plate. It is more difficult to check hip rotation intraoperatively with the patient supine and it is easy to under-correct the medial torsion with a distal osteotomy.

Stable fixation is achieved with both approaches and casting is not required. We find that children mobilize more rapidly after a distal osteotomy but all children should be able to progress to full weight bearing by 3 weeks after surgery.

**SUMMARY**

Single-Event Multi-Level Surgery in spastic diplegia can be considered to be an exercise in correcting anatomical deformities based on clinical and radiological examination and a biomechanical analysis of gait deviations. However, children with spastic diplegia have psychological and physiological dimensions, which make successful surgical outcomes somewhat unpredictable. Weakness is a fundamental issue which is easily overlooked and which may have a greater impact on energy cost of walking and function in the community than multiple musculoskeletal deformities. Cerebral palsy means weakness, originating from causes in the brain. Weakness can be measured using standardized techniques and there is some evidence that strengthening programmes can be successful in improving function. Children and families must be fully prepared for the rigors of multilevel surgery by a careful education program. Choosing the correct age for multilevel surgery is critical but is rarely discussed in the literature. In general terms, the correct age for an individual child is when motor progress plateaus or regresses, because of fixed deformities. In some children the onset of hip subluxation or fixed flexion deformity of the knee heralds the onset of decompensation and that correction of deformities is now urgent. Regular examination of the child is the only means by which the optimum age for multilevel surgery can be chosen. Performing surgery at a very young age leaves many
years of growth remaining during which time deformities may recur. However, leaving surgery to the teenage years is very unwise. Teenagers with physical disabilities have enough to cope with, without a demanding programme of surgery and rehabilitation. In practice, we never perform multilevel surgery before age 6 years. The vast majority of children will have surgery at 7–10 years and we try to have all surgery over before the teenage years.

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