Orthopedic surgery in cerebral palsy: Instructional course lecture

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ABSTRACT
Orthopedic surgery (OS) plays an important role in the management of cerebral palsy (CP). The objectives of OS are to optimize functions and prevent deformity. Newer developments in OS for CP include emphasis on hip surveillance, minimally invasive procedures, use of external fixators instead of plates and screws, better understanding of lever arm dysfunctions (that can only be corrected by bony OS), orthopedic selective spasticity-control surgery, and single-event multilevel lever arm restoration and anti spasticity surgery, which have led to significant improvements in gross motor function and ambulation, especially in spastic quadriplegia, athetosis, and dystonia. The results of OS can be dramatic and life altering for the person with CP and their caregivers if it is performed meticulously by a specialized surgical team, at the appropriate age, for the correct indications, employing sound biomechanical principles and is followed by physician-led, protocol based, intensive, multidisciplinary, institutional rehabilitation, and long term followup. However, OS can be a double-edged sword, and if performed less than optimally, and without the supporting multidisciplinary medical and rehabilitation team, expertise and infrastructure, it often leads to significant functional worsening of the person with CP, including irretrievable loss of previous ambulatory capacity. OS must be integrated into the long term management of the person with CP and should be anticipated and planned at the optimal time and not viewed as a "last resort" intervention or failure of rehabilitation. This instructional course lecture reviews the relevant contemporary principles and techniques of OS in CP.

Key words: Cerebral palsy, orthopedic surgery, orthopedic selective spasticity-control surgery, single event multilevel surgery

MeSH terms: Cerebral palsy, spastic paralysis, encephalopathy, gastrocnemius muscle

INTRODUCTION
Cerebral palsy (CP) is a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to nonprogressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of CP are often accompanied by disturbances of sensation, perception, cognition, communication, and behavior, by epilepsy, and by secondary musculoskeletal problems.1 Although CP is a static encephalopathy, the associated musculoskeletal pathology is often progressive.2 CP affects muscle growth leading to a discrepancy between muscle growth and bone growth. This in turn may lead to deformities of bones and joints, a loss of function, pain, and make care more difficult. These problems get accentuated with increasing age.3 Thus, orthopedic surgery (OS) has a critical role to play in the overall management of a person with CP. As a part of multidisciplinary team, orthopedic surgeons are typically involved in the treatment of spasticity, contractures, joint dislocations, bony deformities and in the overall improvement of musculoskeletal function and ambulation. Pediatric Orthopedic surgeons have pioneered the use of Botulinum toxin followed by serial casting for spasticity management in children with CP over 2 decades ago.4 However, its limitations include high expense, transient
effect lasting a few weeks or months, lack of efficacy in the presence of generalized spasticity, muscle contractures or lever arm dysfunction (LAD), and reports of life threatening complications (permanent paralysis, respiratory paralysis and death in hundreds of children with CP) leading to Food and Drug Administration issuing a warning in 2008 against its use in CP. However, the most crucial function of the orthopedic surgeon in the management of CP is hip surveillance and to ensure no child with CP ever develops a hip dislocation due to delay in ordering a hip X-ray, correctly interpreting it and early surgical correction if indicated. The OS must be goal oriented, address all deformities simultaneously and as early as required. The aim of this instructional course lecture is to review the relevant contemporary principles and techniques of OS in CP.

**Types of Impairments in Cerebral Palsy**

**Primary impairments**

Primary impairments are permanent and are due to the direct effect from the brain lesion, and includes “positive” signs such as spasticity, co-contraction, clonus, hyper-excitability, and released primitive reflex patterns; and “negative” signs such as weakness, fatigability, loss of selective muscle control, poor balance and deficient equilibrium reactions, relative imbalance between muscle agonists and antagonists across joints, sensory problems, and cognitive impairment. The “negative” signs are considered more harmful and disabling than the “positive” signs. In a person who has pure spasticity, only the pyramidal system is damaged; in athetoid CP, only the extrapyramidal system is involved; and in a mixed pattern, both systems are affected. Almost all children with spastic hemiplegia walk independently; most of those with spastic diplegia will walk, often with assistive devices. Those with spastic quadriplegia rarely have functional walking.

**Secondary impairments**

Secondary impairments are due to the after effects of primary impairments, i.e., imbalance between the growth of long bone and musculotendinous units. OS plays an important role in correction of secondary impairments. The effect of spasticity on the growth and development of skeletal muscle results in a muscle that has fewer fibers, shorter fiber length, and a longer tendon. This results in a muscle that is weaker with a diminished excursion because of a decreased cross-section area, resulting in a decreased joint range of motion (ROM). Muscles need the stimulus of stretch to grow. In a child with CP, developmental delay prevents the child from engaging in the typical physical activities that would provide the source of stretch. The hypertonia further impedes the extent of any stretch, restricting the ROM of affected joints and when coupled with musculoskeletal growth, result in “static” muscle contractures where the muscles are both tight and too short. Delays in gross motor skill acquisition and atypical gait patterns lead to further deformity. Common contractures in CP are elbow flexion (biceps and pronator teres) and wrist flexion (forearm flexors) contractures in the upper extremities and hip flexion (psoas), knee flexion (hamstring) and equinus ankle (gastrocnemius) in the lower extremities. The growing bones often become plastically deformed or twisted because of the uneven/abnormal muscle forces present during growth in a person with CP and the spastic muscles cause joint displacement by pulling on the bones. Joint instability in person with CP is often a consequence of both the bony deformity and the unbalanced forces of the agonist-antagonist muscles that bridge across a joint. LAD is the “disruption in the moment generation of a muscle joint complex because of an ineffective lever or moment arm despite normal muscle force.” LAD is also defined as “a set of conditions in which internal and/or external lever arms become distorted because of bony or positional deformities.” Lever is a rigid structure that transmits and modifies force or motion when forces are applied at one point and is able to rotate about another. In the human body, joints function as levers to transmit forces. Lever arm (also known as a moment arm) is the perpendicular distance from the line of application of a force to the axis of joint rotation. Since the muscles and/or ground reaction forces must act on skeletal levers to produce locomotion, LAD greatly interferes with the person with CP’s ability to walk. In a condition, such as CP, the muscle and/or ground reaction forces are neither appropriate nor adequate because of muscle contractures, poor selective motor control, and/or LAD. The types of LAD are: Short lever-arm (coxa valga), flexible lever-arm (pes valgus), malrotated lever-arm (external tibial torsion), an abnormal pivot or action point (hip subluxation or dislocation), and/or positional LAD (crouch gait). The result of LAD is functional weakness and decreased power generation, due to abnormal direction of pull of muscles. LAD becomes clinically significant around the age of 4 years in most cases of spastic diplegia and quadriplegia and then progressively worsens with growth leading to further malalignment of bone as well as unwanted gait compensations. In a recent study, kinematic multilevel rotational anomalies were identified in 98.4% of the 188 children with Spastic Diplegia. LAD does not spontaneously resolve and can only be treated by bony OS, except in mild cases, when bracing can be a temporary option. It is important to understand that muscles cannot be effectively strengthened by physical therapy until co-existing LAD is corrected by bony OS. Early recurrence of contractures is common if LAD is not corrected simultaneously along with surgery of the musculotendinous unit.
Sharan: Orthopedic surgery in cerebral palsy

Tertiary impairments
Tertiary impairments are adaptive mechanisms and coping responses to primary or secondary impairments and must be carefully identified and left alone. An example is circumduction of hip in co-spasticity of rectus femoris and hamstring.6

Natural History of Musculoskeletal Abnormalities in Cerebral Palsy
The natural history in CP is for deterioration in gait and function with time, especially during the growth spurt during adolescence.15 Factors responsible for this worsening may include progression of the musculoskeletal pathology as well as unfavorable changes in the ratio of body mass to strength.16,17 In the lower extremity, the interaction of joint contractures, muscle weakness, co-contraction of agonists and antagonists, bony deformities, and joint instability at multiple levels results in inefficient (higher energy consuming), pathologic and compensatory gait patterns in ambulant children. This leads to limitation in physical function or increase in pain and fatigue. If the child continues walking in the presence of contractures and LAD, the inevitable result is over lengthening of tendons (e.g., calcaneus deformity due to over lengthened tendoachilles) and joint decompensation, after which further ambulation becomes increasingly difficult.18 In nonambulatory children, contractures and deformities of the lower extremities lead to discomfort; difficulties with care, positioning, mobility; and poorer quality of life.11

Requirements for Orthopedic Surgery in Cerebral Palsy
The team of medical and rehabilitation professionals who are going to treat a person with CP must have:
• Knowledge of normal anatomy and physiology, particularly regarding ambulation,
• A good understanding of the functional pathology present in CP,
• Realistic goals/objectives for treatment that are shared commonly by the patient, family, and others concerned with the child’s welfare.
• Knowledge and ability to carry out any of the treatments that are required, and
• A facility with the resources to carry out the necessary evaluations/treatments.6

The minimum requirement for successful conduct of OS is the presence of the orthopedic surgeon or rehabilitation physician-led multidisciplinary rehabilitation team and institutional facilities for an intensive, protocol-based rehabilitation program such as temperature controlled swimming pool for aquatic therapy, body weight supported treadmill training, functional electrical stimulation, virtual reality based therapy, whole body vibration therapy, electromyography (EMG) biofeedback and hippotherapy.5 The rehabilitation professionals must be well trained, experienced, motivated and be prepared to work in a multidisciplinary team under the guidance and supervision of an orthopedic surgeon or rehabilitation physician. Facilities for image intensification and postoperative epidural analgesia (for older children and adults) are recommended. A written and video recorded informed consent with detailed discussion of treatment goals, the time expected to achieve the goals, possible complications, the need for long term followup with the orthopedic surgeon and the possibility of further surgical interventions in the future, with the family is strongly recommended. Though it is important to set reasonable functional goals in discussion with the patient, their caregivers and the rehabilitation team while planning OS, orthopedic surgeons must doggedly avoid offering guarantees regarding functional outcome of OS or attempt to predict improvement in percentage terms, because of ethical considerations and the inability of OS alone to determine the eventual outcome. Orthopedic surgeons must be prepared to personally supervise the postoperative rehabilitation closely, including orthotic management, and followup the person with CP at least till skeletal maturity. The functional outcomes of conventional physiotherapy, especially in an unsupervised domiciliary setting, are significantly inferior to that of physician-led, protocol-based, intensive, multidisciplinary, institutional rehabilitation and patients often become functionally worse after OS in the absence of this rehabilitation.18,17

Timing of Orthopedic Surgery
The timing of OS intervention is determined by CNS maturation, ambulation potential, and the rate at which the contractures and LAD is developing. The presence of significant hip displacement is an absolute indication for OS, irrespective of age. In the absence of hip displacement, one line of thought recommends delaying any OS intervention until ages 7–9 years because of a high risk of recurrence.3 The alternative viewpoint is that the best functional outcomes are achieved between the age group of 4–6 years, before severe LAD and joint decompensation sets in and the risk of recurrence of contractures is actually minimized by simultaneous correction of LAD.5 In spastic diplegia, the formal and intensive OS treatment should be completed between the ages of 4–8 years.19 Although the child develops a mature gait pattern around the age of 7 years,20 it is reasonably close to the mature pattern by the age of 4 years. Once this window of opportunity is lost (usually due to reluctance of other physicians or physiotherapists to let
go or the insistence of the family in exploring nonoperative options at any cost) and complex decompen
dated joint pathology has developed, the results of OS are less gratifying, though functional improvements still occur in older children and adults. Unstable LAD requires OS irrespective of age if there is to be any hope of preserving ambulation. OS must not be viewed as the “last resort to be tried only if everything else has failed. It must be anticipated, planned and carried out at the optimal time by an orthopedic surgeon with subspecialty training in CP surgery, who is backed by a multidisciplinary team and adequate facilities outlined above. OS in adolescence is associated with increased incidence of postoperative psychological problems and sudden disruption of the body image besides longer recovery time due to slower bony healing following osteotomies.

STAGING OF ORTHOPEDIC SURGERY

During the past two decades, correction of all fixed musculoskeletal deformities with single event multilevel surgery (SEMLS) has been the standard of care. SEMLS improves the likelihood of achieving sagittal plane balance and reduces the need for repeated anesthetics, reduces episodes of hospitalization (Mercer Rang’s “birthday syndrome”), and requires only one major period of rehabilitation. It is also more cost effective than staged OS and leads to less disruption of schooling. At present, there are no documented indications for staged OS in CP.

GOALS AND INDICATIONS OF ORTHOPEDIC SURGERY

Goals of OS depend on severity of disease, functional impairment, and level of ambulation (Gross Motor Function Classification System [GMFCS] Level) and the goals of person with CP, family, and the multidisciplinary team. In the first decade of life, the most important priority of persons with CP is function, in the second appearance and in the third and subsequent decades the avoidance of pain. OS should be goal oriented and focus on the projected outcome and quality of life.

The general goals of OS are to:

- Reduce spasticity in a selective manner because an increased muscle tone can sometimes be useful,
- Correct contractures that hinder function or interfere with hygiene,
- Simplify the control system,
- Preserve power generators, and
- Correct LAD and subluxation/dislocation of joints (most commonly the hips).

It is recognized that OS cannot directly address problems with balance, selective motor control, and insufficient strength.

Goals of treatment for ambulatory cerebral palsy (Gross Motor Function Classification System Levels I–III)

- Optimize gait efficiency (correct biomechanics) to optimize energy conservation; (a) preserve or improve physical function, e.g., walk longer distance, walk faster, decrease fatigue, better stability-reduced tripping and fewer falls, keep up with friends; (b) pain relief or pain prevention and increased endurance; (c) preserve or increase activities and participation, e.g., more physically active, more independence, more participation in sports/recreational activity
- Improve appearance of gait: (a) reduced reliance on walking aids; (b) reduced use of orthotics; (c) feet flat on the ground; (d) feet pointing forward; (e) reduced dragging of feet; (f) stand and walk taller (knees less bent); (g) more symmetry

Goals of treatment for nonambulatory cerebral palsy (Gross Motor Function Classification System Levels IV–V)

- Relieve or prevent pain and discomfort
- Facilitate ease of care: Dressing, toileting, bathing/hygiene; positioning: Seating and lying down; transfers and mobility
- Preserve or improve health
- Improve quality of life

PRINCIPLES OF ORTHOPEDIC SURGERY IN CEREBRAL PALSY

The types of OS in CP include release and lengthening of musculotendinous unit, tendon transfers, osteotomies, and arthrodesis. The concept of “surgical dose” in OS for CP, whereby the surgical intervention should match the severity of dysfunction, is relevant.

In CP, multiarticular muscles are more commonly contracted (e.g., psoas, rectus femoris, hamstrings, and gastrocnemii) than the monoarticular muscles. Musculotendinous release or lengthening of monoarticular muscles must be avoided to preserve antigravity function and loss of stability in the postoperative period.

The most common clinical scenario is that of a child with spastic diplegia aged 4–6 years who walks on the toes with the thighs adducted. The usual response of many orthopedic surgeons is to perform tenotomies of adductor longus and brevis (with or without obturator neurectomies) and Z lengthening of the tendoachilles - simple procedures that takes only a few minutes to perform, but the functional consequences for the child may be devastating and permanent. Most children with spastic diplegia have hip and knee flexion contractures, and tiptoe to shift the center
of gravity close to the body. Inappropriate lengthening of tendoachilles in the presence of hip and knee contractures inevitably leads to an unstable calcaneus deformity, which cannot be effectively braced nor salvaged surgically. Besides, the crouch at the knee persists or worsens. It is now evident that what was called “scissoring” gait in the past is actually due to spasticity of medial hamstrings and increased femoral anteversion, in most instances. Adductor tenotomy and obturator neurectomy in this situation will often convert an independent or assisted ambulator into a nonambulator, because of derervation of adductor longus and brevis (important hip flexor and antigravity muscles), besides producing an unacceptable broad-based gait with hyperabduction of the hips.

**Newer Orthopedic Surgery Approaches for Spastic Quadriplegia, Athetosis and Dystonia**

An epidemiological study in India has revealed that the commonest type of CP was spastic quadriplegia (61%) and dyskinetic CP (athetoid and dystonic) accounted for 7.8% of all cases. Orthopedic surgery (OS) is traditionally considered to have a minimal role in this patient population and is even contraindicated by some authors. No other effective treatment option exists for this population, which usually falls under GMFCS Levels IV and V. These persons are at a higher risk of developing hip dislocation, musculoskeletal pain, low bone mineral density and low energy fracture. Prevention of these complications requires that these persons are made ambulant with or without support. However, the recommended rehabilitation strategy at present for these groups across the world is wheelchair aided mobility.

When the musculotendinous unit is lengthened, the muscle belly recoils because of the elastic properties of the musculotendinous unit, it relaxes and assumes a shorter resting length. The muscle is then under less tension, and the spasticity is reduced. As the contracture reduces, the joint ROM increases. Significant decrease in spasticity and both static and dynamic components of muscle tightness was observed following tendon lengthening in children with CP. Decreased spasticity allows the child to have greater ROM, less spastic response to stretch, and better potential to develop and use voluntary muscle activity during gait.

A Japanese orthopedic surgical approach called orthopedic selective spasticity-control surgery (OSSCS) has been proposed with the aim of selective reduction of a specific muscle’s spasticity, dystonia and athetosis, and improvement of antigravity posture control and movement. The principles of OSSCS are:

- Assuming that spasticity of short muscles limits antigravity function in children with CP, longer muscles are selected for release,
- Multiarticular muscles and muscles inserted at the more distal portion in the same muscle group are considered to be the longer muscles,
- The longer and hyperactive muscle fibers can be selectively sectioned with intramuscular tendon lengthening and controlled sliding tendon lengthening,
- Simultaneous release of flexor and extensor muscle groups is performed in each joint (except at wrists, hands and feet, where only the flexor group is released).

The advantages of OSSCS over conventional OS are:

- There is no loss of antigravity activity and weakness of the muscles because monoarticular muscles are preserved,
- Over lengthening of tendons is avoided because of the surgical technique of controlled sliding tendon lengthening,
- It controls spasticity, produces reciprocal movements to facilitate antigravity muscles, and improves functional skills and voluntary movement of the hand,
- It leads to significant functional improvement in the severely involved: Spastic quadriplegia, athetoid, or dystonia
- There is no loss of sensation or sense of stereognosis, and
- There is no increase in the occurrence of dislocations.

Single-event multilevel lever arm restoration and antispasticity surgery (SEMLARASS) is an advancement of the concept of OSSCS. The additional principles of SEMLARASS include:

- Operating between the ages of 4 and 6 years (preferably), to avoid joint decompensation and over lengthening of tendons that happen due to continued usage of deformed joints,
- Simultaneous restoration of LAD is essential for spasticity and contracture correction as well as to reduce chances of recurrence of deformities and repeat surgery at a later stage, and to improve the direction of pull of muscles and thus facilitate strengthening,
- Minimally invasive procedures using image intensification that do not require large skin incisions and consequent risk of blood loss and infection,
- Use of only external fixators that do not require a second operation for removal, and are technically superior to internal fixation with plates and screws in facilitating reduction of dislocated hips using Schanz pins in femoral neck as joysticks, ability to be used in younger children with small bones, and preventing stress shielding of the bone and consequent fractures after removal of plates and screws,
- All surgeries to restore LAD are extra articular to allow for the maximum growth potential of children’s bones,
• Power generators are preserved: Tendon transfers of spastic muscles may lead to further weakness and worse, lead to an opposite deformity, e.g., genu recurvatum following Eggers.
• For nonreducible hip dislocation, the preferred salvage operation is redirection of femoral head and tectoplasty, while preserving the femoral head, and
• The surgery is followed by a structured, intensive, institutional, physician directed, multidisciplinary rehabilitation protocol.

Microinvasive, percutaneous needle tenotomies have been recently described for patients who are not fit for anesthesia.

**ASSESSMENTS AND OUTCOME MEASURES OF CEREBRAL PALSY**

When selecting the assessment methods, the dimensions of the International Classification of Functioning, Disability and Health Child and Youth version should be considered. The following assessment tools and outcome measures are recommended when OS is performed in CP:

**Assessment of functional level and motor development**
The GMFCS is a classification system that allows individuals with CP to be classified according to their level of functional mobility and use of mobility aids. Without reporting GMFCS, it is difficult to generalize the results of OS to the population of children with CP, as the results may only apply to a subpopulation, for example, children with GMFCS Level I–II, who can walk without an assistive device. A systematic review recommended that the tools of choice to measure motor activities of children with CP 12 years and under were Assisting Hand Assessment (AHA), Paediatric Motor Activity Log, Gross Motor Function Measure (GMFM), Gross Motor Performance Measure, and Paediatric Evaluation of Disability Inventory (PEDI). GMFM consists of five different dimensions, and all skills of the children during supine/prone position, sitting, crawling, standing up, and walking is assessed in detail. In addition, the Functional Mobility Scale (FMS) was found to be a clinically feasible tool for quantifying change after SEMLS in children with CP.

**Assessment of muscle tone**
The most frequently used clinical scales are Ashworth/Modified Ashworth Scale (MAS) and Tardieu/Modified Tardieu Scale (MTS). MTS grades muscle spasticity in three different velocities and goniometric measurements are also included for all velocities. MAS is easier to perform and takes less time than MTS, but MTS gives valuable information about muscle length and dynamic contracture and has better intra observer reliability. The commonly used tools to assess dystonia are the Burke-Fahn-Marsden rating scale and Unified Dystonia Rating Scale. The Dyskinesia Impairment Scale is an instrument to measure dystonia and choreoathetosis in dyskinetic CP.

**Assessment of physical fitness**
Activity scales for kids is a commonly used survey, and 6-min walk test is a commonly used measure of general physical endurance.

**Assessment of gait**
The Physician Rating Scale and the Edinburgh Visual Gait Scale are the most commonly used observational gait assessment tools. The Edinburgh Visual Gait Scale has good reliability and concurrent validity.

**Assessment of balance**
Impairments of muscle tone and abnormal postural control in children with CP lead to deficient balance, which is commonly measured by the Paediatric Balance Scale and the Timed Up and Go Test.

**Assessment of trunk impairment**
The Trunk Control Measurement Scale is recommended because it has good inter-rater reliability, only requires basic equipment such as a measuring tape and a ruler, does not require much training, and can be used easily in clinical setting.

**Assessment of health-related quality of life**
The Caregiver Priorities and Child Health Index of Life with Disabilities is recommended due to its strong psychometric properties and clinical utility in children aged 5–12 years with GMFCS Levels III–V.

**Assessment of activities of daily life**
PEDI was reported to be the best assessment instrument for children at an elementary school age because of its psychometric properties and personal ADL items. Children’s Hand-use Experience Questionnaire, Vineland Adaptive Behavior Scale, and Functional Independence Measure for Children were reported to be appropriate for adolescents. Assessment of Motor and Process Skills scale was reported to be the best scale assessing ADL in adolescents with CP regardless of age.

**Assessments of upper extremity**
Manual Ability Classification System (MACS) classifying the upper extremity function at five levels based on bilateral skills of the extremities during daily life activities should be used when reporting the results of OS in the upper extremity. The recommended outcome measures include...
In the upper limb, surgery is also important to improve grasp and release patterns between the wrist and digits, hand appearance, function of the affected hand, and the psychologic status of the patient and the family. The indications for surgery according to Zancolli include:

- Spastic and mixed group with mild athetosis
- Sufficient mental condition and emotional stability
- Low emotional influence on spasticity
- Infantile hemiplegia (especially perinatal)
- Young patients ideally after 6–7 years of age, because training is more effective after this age
- Basic sensibility present, even with some impairment of proprioception and tactile gnosis

Preoperative evaluation should include a thorough physical examination (tone, ROM and strength), use of a functional classification system, and standardized tests that evaluate proprioception, two-point discrimination and stereognosis. Motion analysis and computer modelling of the upper limb are also being increasingly used in some centers.

**Spine Surgery**

The incidence of scoliosis in CP varies from 6% to almost 100%, with the incidence in the overall CP population reported to be 20%–25%. The incidence of scoliosis is the highest in patients with spastic CP (about 70%), in patients with subluxated or dislocated hips (75%) and GMFCS Levels IV and V. The cause of scoliosis in CP is thought to be a combination of muscle weakness, truncal imbalance, and asymmetric tone in paraspinal and intercostal muscles. The natural history of untreated scoliosis in a patient with CP is slow progression from 3 to 10 years of age, rapid progression with pelvic obliquity once curve is over 50° and during the adolescent growth spurt, difficulty with sitting and loss of upper extremity function, increased risk of ischial pressure sores, costo-pelvic impingement and pain, and progression of restrictive lung disease.
Scoliotic curves have been characterized and divided into two groups.63

- Group I: Curves are single thoracic or double thoracic and lumbar curves with level pelvis. This type of curve is typically associated with ambulatory patients (GMFCS II–III).
- Group II: Curves are long thoracolumbar or C-shaped curves with associated pelvic obliquity, typically associated with nonambulatory patients (GMFCS IV–V).

The risk factors for progression of scoliosis in spastic CP include: Having a spinal curve of 40° before age 15 years, having total body involvement, being bedridden, and having a thoracolumbar curve.59 Persons with these risk factors might benefit from early surgical intervention to prevent severe scoliosis. Bracing traditionally has a very limited role in decreasing curve progression.60 Observation is indicated for flexible curves <40° that do not compromise sitting balance. In most cases of severe scoliosis in CP, spinal instrumentation and fusion are recommended because of significant curve progression, loss of sitting balance, and for improved comfort.61 However, spinal deformity correction is a major task in persons with multiple medical co-morbidities and can be associated with a high risk of complications including pulmonary and neurological complications, hardware related issues, infections and death. Expert, multidisciplinary management is required for these challenging conditions.

Surgical site infection is common in children with CP undergoing posterior instrumented fusion for spinal deformity. The incidence is reported to be 1.1%–15.2%. The risk factors include poor nutritional status, patient characteristics (age, cognitive impairment, seizures, presence of ventriculoperitoneal shunts or intrathecal baclofen pumps), and surgery-related factors. The recommended management is irrigation, debridement, and sometimes implant removal.64

**Selective Dorsal Rhizotomy**

Selective dorsal rhizotomy (SDR) is a neurosurgical treatment which is based on a neurosurgical interruption of the afferent of the monosynaptic stretch reflex. The operative procedure usually consists in exposing the dorsal roots L2–S1 and separating them into different rootlets after a laminotomy L2–L5 and opening of the dura. The trans section of the rootlets is performed after electro-stimulation, according to palpable muscle contraction and EMG response. At most, 50% of the rootlets are transected on one level. However, subsequent investigations have demonstrated that the responses to electrical stimulation are inconsistent.65

The selection criteria for SDR is a child with pure spasticity, and affecting predominantly the lower limbs, who could side sit independently, who had some degree of walking ability with good balance and selective motor control, was intelligent and has not undergone prior OS.66 However, a systematic review revealed a complete lack of uniformity in SDR selection procedures around the world.57 SDR is associated with adverse permanent effects (sensory disturbance, bladder incontinence, low back pain, spinal stenosis, scoliosis, lordosis, hip dislocations, and foot deformities), loss of antigravity stability and worsening of motor function, and has no efficacy in patients with contractures, LAD or upper limb involvement.5 More than 10% of patients are likely to develop a spinal deformity after SDR.

The indications of SDR are the same as that for OSSCS or SEMLARASS. Since SDR is extremely invasive, leads to irreversible neural changes, is associated with serious adverse effects and often hastens the need for OS, SDR cannot be recommended at present.

**HIP SURGERY**

The incidence of hip displacement is near 0% for GMFCS Level I but increases to 90% for Level V.68 The hip joint is structurally normal at birth but the spastic muscle imbalance and lack of weight bearing leads to the development of progressive structural changes around the hip joint.69,70 These changes include a relative increase in the neck-shaft angle in the coronal plane (coxa valga), excessive femoral anteversion in the transverse plane, posterolateral acetabular dysplasia, flexion-adduction contractures of psoas and hip adductors, and weakness of hip abductors and extensors.71,72 These deformities contribute to a shift in the mechanical axis away from the center of the femoral head to the lesser trochanter, leading to posterolateral migration of the femoral head and subsequent hip subluxation and dislocation.70 Hip subluxation in early childhood progresses to hip dislocation in 10%–59% of patients by the age of 7–8 years.73–75 If left untreated, hip dislocations may be the source of significant pain due to osteoarthritis and disability in approximately 50% of patients.76 In bilaterally involved children, the muscle contractures and bony deformities may be asymmetric. The more severely involved side might be more adducted and internally rotated. If this is accompanied by relative abduction and/or external rotation of the contralateral side, the deformity is described as “windswept” toward the abducted/externally rotated side.11 This leads to difficulty with dressing, toileting and perineal hygiene. Other potential consequences of a hip dislocation include the development of pressure ulcers, femoral fractures, pelvic
obligation and scoliosis causing problems with sitting. In ambulatory patients, it leads to an energy inefficient gait due to LAD or cessation of ambulatory ability.

The magnitude of the hip displacement is quantified using the Reimer’s Migration Index (RMI). The “hip at risk” (RMI >30%) is defined as the hip that is likely to progress to hip dislocation if left untreated. 3D imaging using computed tomography with 3D reconstruction especially in the direction of the coronal plane are useful in identifying the location and degree of acetabular deformity, assist in preoperative planning and in evaluation of hip pain in the presence of normal X-rays to exclude anterior or posterior dislocation.

The past decade has seen a major interest in the early musculoskeletal screening of children with CP and early OS intervention with the goal being the prevention of extensive or “salvage” surgical procedures that provide sub optimal results. A population-based study using the Norwegian and the Southern Sweden CP registers reported that a surveillance program reduced the prevalence of hip dislocation in the CP population. OS interventions were performed at an earlier age, without increasing the total number of subsequent surgical procedures.

The indications of primary hip surveillance are: Spasticity, delayed walking or inability to walk by 30 months, reduced hip abduction, concern of health-care professional or parent/caregiver. An Australian hip surveillance model recommends annual or bi-annual X-ray of both hips anteroposterior view, as per GMFCS Level and progression of RMI. Monitoring the RMI and acetabular index (AI) are considered the most effective parameters; however, the value of these radiological measurements is limited for displacement directions other than lateral. Children with RMI greater than 33% or AI over 30° are likely to need further treatment. A progression of RMI by more than 7% per annum requires careful monitoring. Based on a recent systematic review, screening is recommended with a single X-ray for GMFCS I and II, or, if RMI >30%, an annual X-ray between ages 2 and 8 years, followed by an X-ray every 2 years until the age of 18 years. For GMFCS III, IV, and V, an annual X-ray is recommended if RMI <30% or 1 every 6 months if RMI >30% between ages 2 and 8 years, followed by X-ray every 2 years until the age of 18 years. With the compelling evidence of the success of a hip surveillance programme, orthopedic surgeons must ensure that no child with CP develops a hip dislocation for lack of timely X-rays and early surgical intervention. Unfortunately, many radiologists are unaware of parameters such as RMI and AI and report on subluxed hip as “normal,” based on which the patient’s caregiver, or their medical/rehabilitation professionals refuse surgery, resulting in an avoidable hip dislocation. Once the child has been included in the followup program, an orthopedic surgeon should be responsible for the hips, arranging radiological screening and the analysis of the results.

The surgical treatment of hip can be grouped as soft tissue (preventive), reconstructive (bony) and palliative (salvage) procedures. Recommended soft tissue surgeries for hip subluxation include psoas recession at pelvic brim and medial hamstring lengthening. As per the principles of OSSCS, release of gracilis, proximal rectus femoris and distal adductor magnus is preferred to tenotomies of adductor longus and brevis. Obturator neurectomy should be strictly avoided. The common clinical practice of performing soft tissue surgery, e.g., adductor tenotomies, at a young age because of the relatively low impact of the procedure on the child and their parents/caregivers is not supported by scientific evidence, if the purpose is prevention or treatment of hip displacement.

One-stage reconstruction of a dislocated hip involves soft tissue procedures, open reduction, and varus derotational (and shortening osteotomies) of the proximal femur (VDRO) to address the angular and rotational deformities. Severe acetabular dysplasia (AI of >25°) in CP can be addressed with surgical procedures redirecting (e.g., Salter osteotomy), reshaping (e.g., Albee, Dega, Pemberton, or San Diego osteotomy) and salvaging/augmenting the depth of the acetabulum (e.g., shelf and Chiari osteotomies). The San Diego periacetabular osteotomy hinges symmetrically on or slightly above the triradiate cartilage, with bone grafts providing posterior coverage equal to the anterior coverage. The Albee, Dega, Pemberton, and San Diego osteotomies reduce the volume and shape of the acetabulum by increasing its lateral coverage without a significant reduction in the posterior coverage.

Unilateral hip bone and soft tissue surgery cause an alteration in the direction of wind sweeping, which causes increased migration of the contralateral hip, and should be avoided. More recently, a minimally invasive, Albee-like percutaneous pelvic osteotomy, VDRO and subtrochanteric osteotomy has been described in nonambulatory patients with hip dislocation. Hip reconstruction is a major event for these vulnerable patients, requires a long period of recovery, can be quite painful, and associated with a substantial risk of complications (femoral fractures, infection, avascular necrosis of femoral head, pressure ulcers, and premature closure of the triradiate cartilage).

Salvage procedures for irreducible hip dislocations include proximal femoral resection (with several variations),
valgus osteotomies, arthrodesis, prosthetic interposition arthroplasty and hip arthroplasty. Salvage procedures, especially femoral head resection, are often followed by chronic, debilitating pain and heterotopic ossification. The author’s preferred salvage procedure is proximal femoral osteotomy to redirect the femoral head and tectoplasty which is consistent with ambulatory capacity and long term pain relief.5,29,30

Pelvic obliquity is a frequent finding and is often associated with scoliosis in a person with severe CP (GMFCS Levels IV and V). Severe pelvic obliquity can affect the ability to sit and cause significant back pain or pain due to rib impingement against the elevated side of the pelvis on the concavity of the curvature. Two patterns of pelvic obliquity have been described: The suprapelvic obliquity is caused by scoliosis, which causes the pelvis to go into obliquity because of extension of scoliosis distally to include the sacrum and the pelvis. Infra pelvic obliquity develops because of fixed contractures in the hip joints, more specifically the windblown hip deformity and subsequent hip imbalance, or subluxation/dislocation of hip joints.91 The infrapelvic obliquity resolves when the windblown hip deformity is corrected. Both these obliquities often co-exist, and progress rapidly during adolescent growth and probably feed into each other, each making the other progress even faster. Surgical correction by spino-pelvic fusion is indicated in persons with progressive deformities which interfere with their level of function and quality of life.91

**LOWER LIMB SURGERY**

Several soft tissue and bony procedures are described to address the musculoskeletal deformities of the knee, foot and ankle in CP with the goal of obtaining an extended and flexible knee, plantigrade, brace able foot, stable base of support for standing and gait, and correction of all LAD.

**KNEE**

The OS procedures include various combinations of musculotendinous lengthening of the distal hamstrings, transfer of the distal hamstrings, lengthening or transfer of the rectus femoris, external rotation and/or extension osteotomy of the distal femur, and distal femoral growth plate (or “guided growth”) surgery.22

The purpose of rectus femoris transfer in a child with stiff-knee gait is to improve the magnitude and timing of swing peak knee flexion that leads to improvements in foot clearance during the swing phase.92,93 The indications of rectus femoris transfer are as follows:

(i) GMFCS Levels I and II (ii) Spasticity of the rectus femoris, confirmed by the Duncan-Ely or prone rectus test (iii) Kinematic variables: A decreased peak knee flexion in swing phase, decreased knee ROM during swing phase, decreased overall knee ROM during the gait cycle, and delay in the timing of peak knee flexion (iv) Dynamic EMG showing prolonged rectus firing during swing phase.92,93

The recommendations of the group from The Royal Children’s Hospital, Melbourne22 regarding OS of knee is as follows:

(i) Medial hamstring lengthening for mild jump gait and <5° knee fixed flexion deformity (FFD) (ii) Medial hamstring lengthening combined with semitendinosus transfer to the adductor tubercle for 5°–15° FFD (iii) Transfer of the semitendinosus combined with growth plate surgery for severe flexed knee gait, combined with knee FFD of 10°–25°, in the children with at least 2 years of growth remaining (iv) Supracondylar extension osteotomy combined with patellar tendon shortening (Ferraretto and Selber technique) for severe crouch gait, combined with knee FFD of 10°–30°, in patients with extensor lag >10°–20° and patella alta on radiographs, who are either close to or already at skeletal maturity (v) Rectus femoris transfer is not performed as a part of SEMLS, but subsequently, if indicated.

However, distal femoral growth plate surgery using staples or 8-plate is yet to show efficacy in long term or controlled studies and compromises the principles of SEMLS in not correcting all deformities in the single surgical sitting. Making a child walk on a knee with persisting FFD could lead to deleterious effects on other joints and lead to over lengthening of tendoachilles.5,29

The concerns with hamstring lengthening include increase in anterior pelvic tilt, loss of hip extension power and ROM. In practice, these effects are usually mild and temporary. Over lengthening of hamstring can lead to genu recurvatum and must be avoided.

The authors preferred approach is OSSCS of medial and/or lateral hamstrings (distal and/or proximal), external rotation osteotomy (to correct femoral anteversion) and/or extension osteotomy of the distal femur (to correct knee FFD), stabilized by external fixator and early mobilization with continuous passive motion. Complete extension of the knee is the goal at the end of surgery. OSSCS of distal rectus femoris (instead of transfer) is simultaneously added for stiff knee gait.5

**ANKLE AND FOOT**

The commonest foot segmental malalignments in persons with CP are equinus, equinoplanovalgus, and
equinocavovarus. These deformities are often associated with ankle valgus and hallux valgus. Foot and ankle deformities caused by dynamic over activity and muscle imbalance are best treated with pharmacologic or neurosurgical interventions for spasticity, or muscle tendon unit transfers. Deformities caused by fixed or myostatic soft tissue imbalance without fixed skeletal malalignment are best treated with musculotendinous unit lengthening surgery. Deformities characterized by structural skeletal malalignment associated with fixed or myostatic soft tissue imbalance are best treated with a combination of soft tissue and bony surgeries. Bony surgeries include osteotomies and arthrodesis, and can correct deformity by addition (i.e., lengthening), subtraction (i.e., shortening), angulation, or rotation. Osteotomy is always preferred to arthrodesis to restore foot skeletal segmental alignment and maintain intra- and inter-segmental motion, to optimize shock absorption and lever functions of the foot.

The most common deformity is equinus. In the presence of significant contractures of the gastrocnemius and/or soleus, if the foot is developing a severe midfoot break or the child can no longer tolerate the plantigrade orthotics, surgical lengthening is indicated. Children with spastic CP and equinus gait have longer than normal tendoachilles and shorter than normal muscle bellies. The surgical lengthening of muscle tendon unit should specifically focus only on the contracted muscle, in diplegia typically only the gastrocnemius, with a mid calf myofascial release. In hemiplegia, the soleus also sometimes develops contractures and a combined slightly more distal myofascial release of the conjoined tendon is preferred. It must be emphasized that it is preferable to have several recurrent equinus contractures than to have one overcorrection.

The planovalgus deformity is next commonest deformity, and begins with the lateral displacement of the navicular, causing the talar head to become uncovered and prominent in the medial midfoot. OS for planovalgus is indicated if the foot is painful during gait or the mechanical stability of the foot is impacting the ability to achieve optimal gait. Lateral calcaneal lengthening with concomitant peroneus brevis lengthening is the recommended procedure for correcting a planovalgus foot deformity.

For ankle valgus in children with open physes and more than 2 years remaining growth, reversible hemiepiphyseodesis using a medial malleolar screw is preferred. Medial displacement calcaneal sliding osteotomy is the preferred surgery for hind foot valgus.

Severe rigid equinocavovarus deformity can be treated with a one stage correction including soft tissue surgery, lateral displacement calcaneal sliding osteotomy and/or lateral closing wedge calcaneal osteotomy, a midtarsal dome osteotomy and a modified Jones procedure for a first metatarsal drop.

The recommended surgical techniques for hallux valgus include first metatarsophalangeal joint arthrodesis, or proximal first metatarsal osteotomy, distal soft tissue releases, and exostectomy of the bunion. The most common deformity is equinus. In the presence of significant contractures of the gastrocnemius and/or soleus, if the foot is developing a severe midfoot break or the child can no longer tolerate the plantigrade orthotics, surgical lengthening is indicated. Children with spastic CP and equinus gait have longer than normal tendoachilles and shorter than normal muscle bellies. The surgical lengthening of muscle tendon unit should specifically focus only on the contracted muscle, in diplegia typically only the gastrocnemius, with a mid calf myofascial release. In hemiplegia, the soleus also sometimes develops contractures and a combined slightly more distal myofascial release of the conjoined tendon is preferred. It must be emphasized that it is preferable to have several recurrent equinus contractures than to have one overcorrection.

Outcome of Orthopedic Surgery

SEMLS results in clinically and statistically significant improvements in gait and function, in children with bilateral spastic CP, which were maintained at 5 years after surgery, as per a Randomized Controlled Trial. In a systematic review of SEMLS, large improvements in gait were noted in most studies reviewed. The systematic review also reported rather small changes in gross motor function. No studies, which included both summary statistics of gait and GMFM, showed a significant improvement in both. Changes in function as measured by the GMFM66 showed an improvement of 3.3% at 5 years compared to presurgery. When it is considered that the natural history of gross motor function
function in this age group is for deterioration, this finding becomes even more important and clinically significant.\textsuperscript{105}

The deformity correction is usually immediate after SEMLS. However, clinically and statistically significant improvements in gait were found only 12 months after SEMLS and functional improvements (GMFM66) were not found until 2 years after SEMLS.\textsuperscript{106} Studies on ambulatory children at GMFCS Level II and III have shown that the GMFCS remains unchanged or “stable” in 95% of children following SEMLS and improves by 1 level in 5%.\textsuperscript{107}

Functional results of 314 persons who underwent SEMRARASS showed median value of FMS of 3 before surgery and 5 after surgery. Before surgery the median value of GMFCS was Level IV and after surgery it was Level II. The GMFCS improved 2 levels on average. Before surgery, median value of MACS was III and after surgery it was I. A significant improvement was noted in participation levels, motivation and a significant improvement in the overall quality of life. Persons with severe CP (GMFCS IV and V) showed more positive correlation than mild to moderate cases.\textsuperscript{5}

**Complications of Orthopedic Surgery**

The intraoperative complications include intraoperative hypothermia (incidence 26.2%), absolute hypotension (4.4%), and absolute bradycardia (20.0%). Intraoperative and major immediate postoperative complications were significantly higher in patients at GMFCS Levels IV and V, older age, hip reconstructive surgery, and history of pneumonia.\textsuperscript{108} 2.3% patients had major complications, and 4.0% patients had minor complications postoperatively.\textsuperscript{108} Other perioperative complications include fat embolism, cardiorespiratory and gastrointestinal complications, wound infection and wound bleeding. Complications of OS can be classified into under correction (e.g., inability to achieve a horizontal pelvis after posterior instrumentation and fusion for thoracolumbar scoliosis), overcorrection (e.g., a calcaneovalgus deformity after tibialis anterior transfer for equinovarus deformity of foot) or recurrence following a successfully treated deformity. Recurrence is common after OS in CP and is usually due to failure to assess and correct LAD and all concurrent deformities, inadequate/ineffective rehabilitation and orthotic management, and loss to followup.

Complications encountered during rehabilitation following SEMRARASS in 463 consecutive patients included myofascial pain syndrome (149, 32.60%), prolonged articular stiffness beyond 4 weeks (111, 24.23%), patellofemoral pain syndrome (38, 8.13%), osteopenia (36, 7.88%), meralgia paresthetica (26, 5.69%), pressure ulcers (19, 4.10%), hypertrophic scar (18, 3.94%), low energy fractures (19, 4.06%), superficial pin tract infection (12, 2.56%), wound dehiscence (9.1.92%), patellar tendinosis (8, 1.71%), and myositis ossificans (7, 1.51%). A preoperative GMFCS Level IV and V was associated with a higher prevalence of complications like osteopenia, low energy fractures and myositis ossificans. However, none of the complications were life threatening, permanent or affecting the long term outcome of surgery. To minimize the rate of complications a structured rehabilitation protocol carried out by an experienced multidisciplinary medical team was recommended.\textsuperscript{5}

**Perioperative Management**

The same multidisciplinary team is involved in the perioperative management, with the additional and essential support of the experienced anesthetist and nurses. Effective pain control is essential and allows early postoperative mobilization and discharge. Careful preoperative assessment and optimal management of comorbidities is required and benefit/risk ratio of each OS intervention considered preoperatively.

**Followup**

The number of followup visits and the duration of followup depends on the age of patient, nature of OS, the level of severity of the CP (GMFCS Level), presence of complications and comorbidities, need for hip surveillance and the available community resources. Objective outcome assessment is essential during followup visits. Followup at least till skeletal maturity is essential.

**Factors Affecting outcome of Orthopedic Surgery**

The following factors have been found to determine the final functional results following SEMRARASS:

- Dysfunction of lever arm in the lower extremities
- Epileptic episodes in last 2 years
- Excessively lengthened tendons (e.g., due to previous botulinum toxin injections or tendon lengthening surgery)
- Postoperative complications in relation with severity and its duration
- Abnormalities of bone mineralization (low bone mineral density or Vitamin D)
- Known cognitive deficits
- Sensory problems, e.g., visual or auditory impairments
- Hand function
- Age at the time of surgery
Sharan: Orthopedic surgery in cerebral palsy

- Rehabilitation in terms of intensity, quality and duration
- Achieved level of gross motor function at the time of surgery
- Neurosurgery (rhizotomies, fasciculotomies) in the past
- Socioeconomic background.

CONCLUSION

OS intervention in a person with CP is a process rather than an isolated event which has the potential to significantly improve function in the short term and reduce the burden of care in the long term. A continuing improvement in the functional outcome of OS intervention in persons with CP is only possible in the presence of meticulous preoperative assessment and preparation, expert perioperative management and long term followup, within the setting of a multidisciplinary service and in partnership with the person with CP and family. OS that simultaneously and effectively addresses spasticity, abnormal movement patterns and LAD and followed by intensive, physician-led protocol-based functional rehabilitation and long term followup provides the person with CP the best functional outcome.

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