The HELP Guide To
Cerebral Palsy
Second Edition

Authors
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Consultants
Leon ROOT
Lynn STAHELI

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GLOBAL HELP
Health Education using Low-cost Publications

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Preface

Drs. Nadire Berker and Selim Yalçın created a second edition of their successful publication, The Help Guide to Cerebral Palsy. This publication is authoritative, comprehensive, extensively illustrated, colorful and engaging. The need for this publication is enormous, as CP is common throughout the world and no affordable management guide has been available to date.

The authors are highly qualified to produce this publication. Dr. Nadire Berker was Professor of Rehabilitation Medicine at the Marmara School of Medicine in Istanbul, Turkey. She has extensive experience in managing the full spectrum of CP from childhood to adult life. Dr. Berker continues a family tradition in rehabilitation medicine that was started in the 1920s by her grandfather who established the specialty in Turkey, and was continued by her mother who headed the department for many years. Dr. Selim Yalcin is Professor of Orthopedics at the same university. Dr. Yalcin is an accomplished clinician who specializes in pediatric orthopedics. He has broad clinical experience, and is a prolific author and producer.

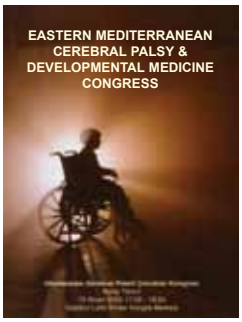
The authors have produced numerous books and videos on cerebral palsy, spina bifida, gait analysis, clubfoot, the use of ultrasonography in orthopedic management, and historical aspects of medicine.

They practice medicine in Istanbul, a city positioned at the crossroads of the world. The economy of Turkey is midway in the economic spectrum of nations of the world. This geography and economic environment provides the authors with an ideal vantage point from which to make management recommendations that are both practical and relevant for most societies of the world.

The authors recommend a balanced approach to management. This approach balances the medical, social, psychological and educational needs of the child and family. The humanitarian, whole-child approach is evident throughout the publication.

Global HELP Organization is pleased to support The HELP Guide to Cerebral Palsy. The book is available through our web site in pdf format; for non-commercial use, it may be downloaded without charge. Printed copies are available for only the cost of printing and mailing. Please visit our web site at www.global-help.org for details.

.Lynn Staheli, MD, 2010
Founder and Director
Global HELP Organization
Seattle, WA, USA



The Story of the EMCPDM

The authors of this book were among the group of dedicated American Academy of Cerebral Palsy and Developmental Medicine (AACPDM) members who felt the necessity to improve the quality of health care services urgently needed by children with neuromuscular diseases in the Middle East. After long discussions on what to do, they planned to start meetings to provide a regional platform for educating physicians and therapists. Drs. Berker and Yalçın shouldered the responsibility of the first meeting in Istanbul, performing the secretarial work, graphic design, and fund raising by themselves. With seed money donated by the AACPDM, they organized a large meeting at which recent scientific advances in the area of childhood neuromuscular disability and CP were the main topics.

The first Eastern Mediterranean Cerebral Palsy and Developmental Medicine meeting was held in 2002 in Istanbul. More than 300 participants from 18 countries were in attendance. For the 3-day congress in the most modern convention center in Turkey, registration fees were kept to a minimum of $100 and free accommodation was provided to participants from countries with economical problems.

The faculty included renowned physicians and physiotherapists from the United States, Europe and the Middle East, most of whom volunteered to participate and donated their time and efforts. The success of the first meeting led to the next in 2004 in Greece, which also was very successful. Biannual meetings continued in 2006 in Warsaw, Poland; in 2008 in Eilat, Israel and in 2010 Beirut, Lebanon.

The organization today covers Eastern Europe as well as the Middle East, and therefore is called the EEMCPDM (Eastern European and Mediterranean Group of Cerebral Palsy and Developmental Medicine). Over a period of ten years, it has evolved into a wonderful opportunity for the physicians and therapists of the region to get together, share knowledge and experience, and discuss the latest developments. The group is looking forward to future meetings with proud anticipation.

Dear Reader,

Years of treating children with cerebral palsy (CP) has shown us that a worldwide need exists for a concise, illustrated book to guide health professionals regarding this difficult problem. This book is an attempt to fulfill that need. The *Guide* is intended for use by physicians, residents, medical students and allied health professionals who treat children with CP. We focus on the latest concepts in the treatment of musculoskeletal problems and describe the associated impairments, providing suggestions for further reading. The chapters on total body involvement, diplegia, hemiplegia and dyskinesia include the most common treatments applied for these patients. Note that problems described in one section may occur in other types of CP. We present the most frequently used and accepted treatment methods with scientifically proved efficacy and include references at the end of each chapter.

The illustrations and photographs of patients are from ours and Dr. Leon Root's archives unless stated otherwise. We would like to thank our patients and their families for sharing their problems with us and also for allowing us to use their pictures. We are indebted to Ms. Dory Kelly for helping us with text editing of the first edition.

Treatment of the child with CP is difficult, often frustrating and sometimes depressing. This is even more pronounced in countries with limited resources for the disabled. We try to provide information on how to proceed in places where resources are limited. An interdisciplinary management approach is the only means to integrate children with CP with the society and lessen the impact of the problem. We have learned with great pleasure that ten thousands of readers benefited from our work and used this guidebook in the treatment of unfortunate millions of patients with CP worldwide. Therefore we thought it necessary to update the guide with knowledge and expertise of the last five years. We hope it will be of valuable use to all persons involved in taking care of children with cerebral palsy.

Nadire Berker and Selim Yalçın
Istanbul, Turkey - 2010

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Leon Root, MD

Dr. Root is one of the leading names in cerebral palsy orthopaedic surgery worldwide. A former president of the AACPDM, he has dedicated his career to cerebral palsy, given many lectures and authored more than 100 articles on the subject.

Lynn Staheli, MD

Dr. Staheli is one of the most prominent names in pediatric orthopaedics. He is the founder and chief editor of the Journal of Pediatric Orthopaedics, author of six books and a wizard of desktop publishing. He founded the Global HELP organization to create affordable medical textbooks worldwide.

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Lana Staheli, PhD is a certified counselor, marriage therapist, and consultant on interpersonal relationships and life strategies. She authored several relationship books. Lana is co-founder and vice-president of Global HELP.

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Dr. Koloyan is a pediatric orthopaedic surgeon from Yerevan, Armenia. He has been one of the creators of the EMCPDM and has done pioneer work for disabled children of Armenia and Georgia.

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Dr Papavassiliou is the director of Pediatric Neurology at the Pendeli Children's Hospital in Athens, Greece. She has been treating children and adolescents with CP for many years and has devoted a lot of time in teaching physicians and therapists. She has co-authored two medical textbooks on CP and many others for patient education in Greek.

Idil Çilingiroğlu

Ms. Çilingiroğlu is an architect who devoted her time and talent to draw all the illustrations in the section for families.

Foreword

Cerebral palsy (CP) is the most common chronic disability of childhood today. It is ubiqitious and it occurs all around the world. In developed nations, the incidence is about 1 to2 per 1000 births. In spite of improved obstetrical and perinatal care, CP remains with us. As a result of injury to the brain, these children have motor defects which will affect them for their entire lifetime. Treatment often starts when they are infants, and continues throughout their life, even into adulthood. The problems involved are complex; not only do these children have problems of mobility, but they can also have seizure disorders, gastrointestinal system problems, learning and perceptual difficulties, visual problems, hearing problems, and growth deficiency. In spite of all these numerous difficulties, cerebral palsied children can be helped.

What the authors attempt in this book is to divide information for physicians, therapists or other paramedical personnel who are interested and will be treating and taking care of these children for their lifetime. They present the basic understanding of what CP is and the fact that it takes a team to treat them. The child and his parents become the focus of treatment because you cannot treat the child without involving the parents as well. The team has to consist of the physician who will be the captain of the team. That physician can be a pediatrician, orthopaedic surgeon, physiatrist or even a neurologist, but they must take overall control of the child and make sure that all parameters of care are attended to. The team has to also include the occupational therapist, the speech therapist, the physical therapist, teachers and social workers.

Dr. Wallace Salter of Toronto, Canada is fond of saying that after you operate on the child with CP, he still has CP. This may seem discouraging because we cannot cure the CP with our present knowledge, but we can make life better for these children, and that is important. Even small degrees of improvement makes a great difference. Getting a child to walk, be it in crutches, in braces or with a walker, is much better than having him in a wheelchair. Having a child be able to live in a wheelchair, as is true for children with total body involvement, is much better than having him be on a stretcher or in a bed for the rest of his life. These are important things to consider.

The authors have carefully defined the types of CP, the prognosis of CP, therapies that are at present available, the surgical indications and most important of all, the pre and postoperative care that these children must have. They write in a very clear and concise way which provides a ready reference for the interested reader in treating these children.

In my own experience, I have found that working with the cerebral palsied children and their families has been the most rewarding aspect of my medical career. The children and families are deeply grateful to you for whatever you can offer them and particularly, they respond to the fact that you care. And it's with your caring and your ability to help that makes a difference.

Leon Root, MD
Orthopaedic Surgeon
Hospital for Special Surgery
New York, February 2005

Dedication

We would like to thank

Our mothers and fathers, for their guidance and inspiration.


Asaf Yalçın


Sabahat Yalçın


Ender Berker


Mustafa Berker

Our children, for their compassion towards those less fortunate than themselves.


Deniz Yalçın


Deniz Özaras


Güneş Yalçın

General Concepts

Definition

Cerebral Palsy (CP) is a disorder of movement and posture that appears during infancy or early childhood. It is caused by nonprogressive damage to the brain before, during, or shortly after birth. CP is not a single disease but a name given to a wide variety of static neuromotor impairment syndromes occurring secondary to a lesion in the developing brain [A]. The damage to the brain is permanent and cannot be cured but the consequences can be minimized [B]. Progressive musculoskeletal pathology occurs in most affected children.

The lesion in the brain may occur during the prenatal, perinatal, or postnatal periods [C]. Any nonprogressive central nervous system (CNS) injury occurring during the first 2 years of life is considered to be CP.

In addition to movement and balance disorders, patients might experience other manifestations of cerebral dysfunction [D].

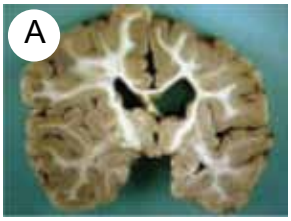
CP was first described by the English physician Sir Francis William Little [E] in 1861 and was known as *Little's disease* for a long time. Little thought that this condition was caused by neonatal asphyxia [F]. Later, Sigmund Freud [G] and other scientists challenged Little's idea and proposed that a variety of insults during pregnancy could damage the developing brain. Today, it is accepted that only approximately 10% of cases of CP can be attributed to neonatal asphyxia. The majority occur during the prenatal period, and in most of the cases, a specific cause cannot be identified.

Epidemiology

CP is the most common cause of childhood disability in Western societies. The incidence is 2-2.5/1000 live births. Some affected children do not survive and the prevalence varies between 1-5/1000 babies in different countries. It was previously thought that improvements in perinatal and obstetric care would decrease the incidence of CP. However, the incidence has not declined and the overall prevalence increased during the 1980s and 1990s. This is explained by increased survival of premature and very-low-birth-weight infants and by a rise in the number of multiple births. Even at centers where optimal conditions exist for perinatal care and birth asphyxia is relatively uncommon, the incidence of CP in term babies has remained the same. This has led researchers to consider unknown prenatal causative factors.

Etiology

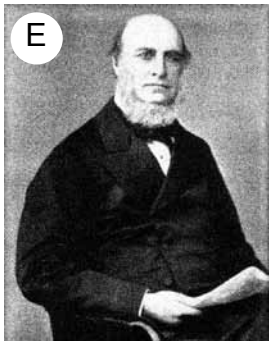
The etiology can be identified only in 50% of the cases. Certain factors in the history of the child increase the risk of CP. The incidence of CP among babies who have one or more of these risk factors is higher than among the normal population. The clinician should therefore be alerted to the possibility of the presence of CP in a patient with these factors.



The brain lesion cannot be cured but it does not get worse either.

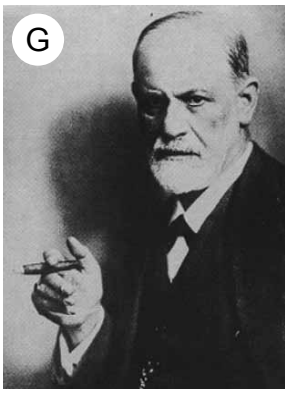
Time of brain injury	C
Prenatal period	Conception to the onset of labor
Perinatal period	28 weeks intrauterine to 7 days postnatal
Postnatal period	First two years of life

Manifestations of cerebral palsy	D
Neurological	*Associated problems*
Muscle weakness	Intellectual impairment
Abnormal muscle tone	Epilepsy
Balance problems	Visual problems
Loss of selective control	Hearing loss
Pathological reflexes	Speech and communication problems
Loss of sensation	Swallowing difficulty
Musculoskeletal	Feeding difficulty, failure to thrive
Contractures	Respiratory problems
Deformities	Incontinence



In 1861 William Little described a paralytic condition of children that is recognizable as spastic diplegia.

In his paper, Dr. Little showed a child with adductor spasm, crouch gait and intoeing.



Sigmund Freud challenged Little's opinion on the association between CP and birth trauma.

"Cerebral palsy (CP) describes a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication, and behaviour, by epilepsy, and by secondary musculoskeletal problems."Rosenbaum et al, 2007

CP has been defined as a non progressive injury to the immature brain leading to motor dysfunction. Although the lesion is not progressive, the clinical manfestations change over time (Mercer Rang).

Risk factors (A)
Prenatal
Prematurity (gestational age less than 36 weeks)
Low birth weight (less than 2500 g)
Maternal epilepsy
Hyperthyroidism
Infections (TORCH)
Bleeding in the third trimester
Incompetent cervix
Severe toxemia, eclampsia
Hyperthyroidism
Drug abuse
Trauma
Multiple pregnancies
Placental insufficiency
Perinatal
Prolonged and difficult labor
Premature rupture of membranes
Presentation anomalies
Vaginal bleeding at the time of admission for labor
Bradycardia
Hypoxia
Postnatal (0-2 years)
CNS infection (encephalitis, meningitis)
Hypoxia
Seizures
Coagulopathies
Neonatal hyperbilirubinemia
Head trauma

Risk factors

Risk factors associated with CP are grouped into prenatal, perinatal, and postnatal factors [A]. Prematurity and low birth weight are the two most important risk factors in developed countries with high standards of obstetrical care. Postnatal risk factors additionally play a major role in other countries.

A clear association exists between premature delivery and spastic diplegia [B]. Low birth weight increases the risk. Rubella, herpes simplex, toxoplasma, and cytomegaloviruses cross the placenta to infect the fetus and have severe effects on the developing CNS. Eclampsia or other severe maternal illness hypothermia, hypoglycemia of the neonate cause a reduction in the levels of oxygen and nutrients available to the fetus or an increase in the levels of toxins or waste products, adversely affecting the developing CNS. Multiple pregnancies or breech presentation also can increase the risk [C]. Excess of bilirubin resulting from the haemolytic disease of the newborn is clearly associated with CP.

Babies who carry these risk factors should be under close supervision by a pediatric neurologist for signs suggestive of neuromotor developmental delay.

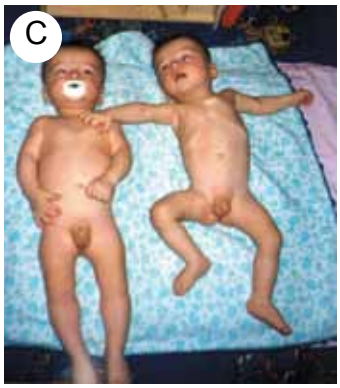
Pathological findings in the CNS

Specific brain lesions related to CP can be identified in most of the cases. These lesions occur in regions that are particularly sensitive to disturbances in blood supply and are grouped under the term *hypoxic ischemic encephalopathy*.

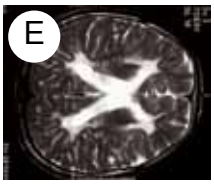
Five types of hypoxic ischemic encephalopathy exist [D]; parasagittal cerebral injury, periventricular leukomalacia [E], focal and multifocal ischemic brain necrosis [F], status marmoratus and selective neuronal necrosis.


B

Approximately 11% of premature babies who survive in neonatal intensive care units develop CP.


C

In vitro fertilization results in multiple pregnancies and increases the risk of CP.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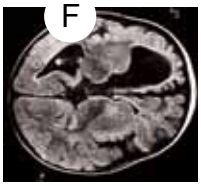
Periventricular leukomalacia (PVL)


F

Multifocal ischemic brain necrosis

Subtypes of hypoxic ischemic encephalopathy (D)		
Lesion	*Location*	*Clinical Finding*
Parasagittal cerebral injury	Bilateral in superior medial and posterior portions of the cortex	Upper extremities more severely affected than lowers
Periventricular leukomalacia	Bilateral white matter necrosis near lateral ventricles descending fibers of the motor cortex, optic and acoustic radiations	Spastic diplegia and quadriplegia visual and cognitive deficits
Focal and multifocal ischemic brain necrosis	Infarction in a specific vascular distribution (most commonly left middle cerebral artery)	Hemiplegia and seizures
Status marmoratus	Neuronal injury in the basal ganglia	Choreoathetosis or mixed
Selective neuronal necrosis (usually combined with the above)	Lateral geniculate, thalamus and the basal ganglia	Mental retardation, seizures

Clinical findings

Children with CP present with three types of motor problems [A]. The primary impairments of muscle tone, balance, strength and selectivity are directly related to the damage in the CNS. Secondary impairments of muscle contractures and deformities develop over time in response to the primary problems and musculoskeletal growth. Tertiary impairments are adaptive mechanisms and coping responses that the child develops to adapt to the primary and secondary problems. One typical example is gastrocnemius spasticity as a primary impairment [B] leading to secondary ankle plantar flexion contracture and knee hyperextension in stance [C] as an adaptive mechanism.

Mechanism of the movement problems

Abnormal muscle tone, disturbance of balance mechanisms, muscle weakness and loss of selective motor control lead to an inability to stretch muscles. Muscle weakness, spasticity, and contractures [D] also result in abnormal skeletal forces which cause bone deformity [E] as the child grows older.

Muscles grow through stretch. Stretch occurs during active movement. When the child wants to play, he moves and stretches the muscles. This creates the necessary input for muscle growth. The child with CP cannot play because of pathological tone, weakness, poor selective control and abnormal balance. His muscles are not stretched and do not grow. The distal biarticular muscles are more affected because selective motor control is worse distally and the biarticular muscles [F] are more abnormal than are the monoarticular muscles.

The child with CP has abnormalities of muscle tone and reflexes, shows delay in developmental milestones, and presents with posture and movement problems. When he tries to move, muscle contractions cannot be effectively controlled. This is a result of many factors [G].

A

Primary impairments
(due to the brain lesion)
Muscle tone (spasticity, dystonia)
Balance
Strength
Selectivity
Sensation
Secondary impairments
(due to the primary impairments causing the movement disorder)
Contractures (equinus, adduction)
Deformities (scoliosis)
Tertiary impairments
Adaptive mechanisms (knee hyperextension in stance)

Equinus contracture due to triceps surae spasticity is a secondary impairment.

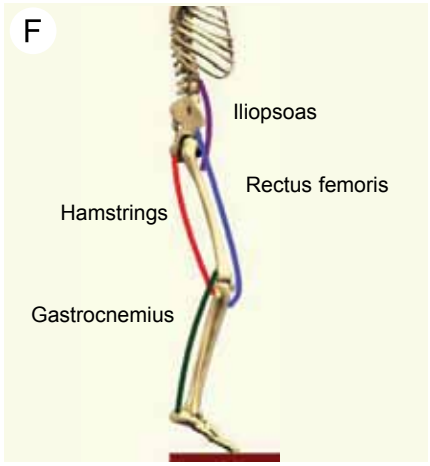
Knee hyperextension is an adaptive response to equinus deformity.

D

Common sites for contracture	
Upper extremity	*Lower extremity*
Pronator	Hip adductor-flexor
Wrist and finger flexor	Knee flexor
Thumb adductor	Ankle plantar flexor

E

Common sites for deformity	
Spine	Scoliosis, kyphosis
Hip	Subluxation, dislocation
Femur & tibia	Internal or external torsion
Foot	Equinus, valgus, varus



Biarticular muscles of the lower extremity are most commonly involved.

G

Causes of the motor problem
The muscles are weak and cannot generate the appropriate force necessary for movement.
Spasticity does not allow the muscle to relax. It causes unnecessary contractions during movement.
The coordinated contraction and relaxation of many muscles is necessary for a smooth movement. Certain muscles need to relax while others contract. The cerebral centers controlling this complex selective motor control are disturbed in CP. The child is unable to relax certain antagonist muscles and contract the agonists necessary for a specific task.
Primitive reflexes interfere with the development of gross and fine motor control.
Advanced postural reactions for balance and equilibrium that are a prerequisite for sitting and walking are either delayed or nonexistent. When the child cannot sustain balance, movement becomes more difficult.
Apraxia (inability to plan and execute motor function) is present.
Superficial sensation is generally normal, cortical sensation, proprioception and sensation of movement may be impaired.

Maturation of the central nervous system
Primitive reflexes
Cutaneous
Palmar grasp
Plantar grasp
Rooting
Sucking
Gallant
Labyrinthine
Prone
Supine
Proprioceptive
Symmetric tonic neck reflex
Asymmetric tonic neck reflex
Moro
Foot-hand placement
Advanced (postural/protective) reactions
Head righting
Head and body righting
Protective-antigravity
Forward-lateral-backward reactions
Parachute-protective extension response
Landau
Equilibrium reactions
Voluntary movement
Fine motor
Gross motor
Rolling
Sitting
Standing
Sphincter control

A

Evolution of CP during infancy and early childhood

The movement problem associated with CP is not apparent in infancy. It is established during the period of CNS development. Children who are going to have CP show neuromotor developmental delay in infancy. The typical clinical picture is established toward the age of 1 year in a number of these children. Movements become normal as the nervous system matures in some others.

The normal newborn demonstrates primitive reflex movements. These are complex, stereotypical patterns that occur in response to a variety of sensory stimuli. At birth almost all motor behavior is controlled by these primitive reflexes. Within a few months, they are replaced by a more mature set of protective and postural reflexes called advanced postural reactions that position the body segments against each other and gravity. Advanced postural reactions provide the basis for trunk balance and voluntary control of movements. The child gains motor skills as primitive reflexes are supressed and advanced postural reactions are established [A].

Primitive reflexes persist [B-D] and advanced postural reactions [E,F] do not appear in the child with CP. Abnormal movement patterns emerge as the child grows.

The child's ability to achieve head control, sit, crawl, stand, and walk

D

Tonic labyrinthine reflex: Put the baby in the prone position, bring the neck to 45º flexion. The response is flexion of the arms, trunk and legs. Put the baby in the supine position and bring the neck to 45º extension. Extensor tone will increase. This reflex is present at birth and disappears at 4 months.

B

Asymmetrical tonic neck reflex (ASTNR): The child lies supine. Turn the head to one side and then the other. The extremities on the face side extend and the ones on the occiput side flex. This is called the fencing position. The reflex is present at birth and disappears at 6 months.

Photo courtesy of G. Koloyan

C

Foot placement reaction: Hold the child by the axilla and bring the dorsum of the foot against the edge of the table. The child will automatically place his foot on the table top. This is a normal response in all children and is inhibited by age 3 to 4.

E

Landau reflex: Suspend the baby horizontally. The neck, trunk and arms extend, legs partially flex. This is an advanced postural reaction which appears at 6 months.

F

Parachute response: The child lies prone on the table. Lift the child vertically and suddenly tilt forward towards the table. The arms and the legs extend as a protective reaction. This is an advanced postural reaction that appears at 8 months.

is always delayed. Late achievement of a milestone such as sitting indicates the presence of a motor deficit and the degree of delay correlates with the severity of the problem [A,B].

Babies with CP usually have a period of hypotonicity during the early months of life. Between the ages of 6 to 18 months, muscle tone gradually increases in those who are going to develop spasticity. Fluctuations in tone from hypo- to hypertonicity is a characteristic of developing dyskinetic CP. Athetosis becomes obvious after 18 to 24 months. Ataxia may not be apparent until even later.

Early signs suggestive of CP in the infant are abnormal behavior, oromotor problems and poor mobility [C]. The infant is irritable, too docile, or difficult to handle. He does not suck well, sleeps poorly, vomits frequently and has poor eye contact. Deviant oromotor patterns include tongue retraction and thrust, tonic bite and grimacing. Early motor signs are poor head control [D] with normal or increased tone in the limbs [E], and persistent or asymmetric fisting. Motor development is both delayed and abnormal [F]. Instead of crawling, the child moves by creeping or hopping like a bunny. Hand preference during the first two years of life is a sign of hemiplegic CP.

The clinical picture of CP is established in early childhood as the movement problem becomes prominent [G,H].

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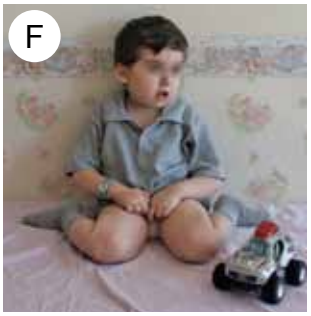


Children with increased femoral anteversion and adductor spasticity sit in the W-position to maintain balance.

Pathological asymmetrical posture in a 6 year old child

Absent Landau reflex at 11 months is a sign of developmental delay.

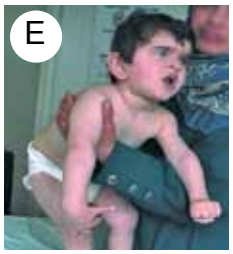
B **CP is likely if there is no**

Head control	3 months
Sitting	6 months
Rolling over	6 months
Walking	18 months

C **Signs suggestive of CP in an infant**

- Abnormal behavior
  - Excessive docility or irritability
  - Poor eye contact
  - Poor sleep
- Oromotor problems
  - Frequent vomiting
  - Poor sucking
  - Tongue retraction
  - Persistent bite
  - Grimacing
- Poor mobility
  - Poor head control
  - Hand preference before 2 years of age
  - Abnormal tone

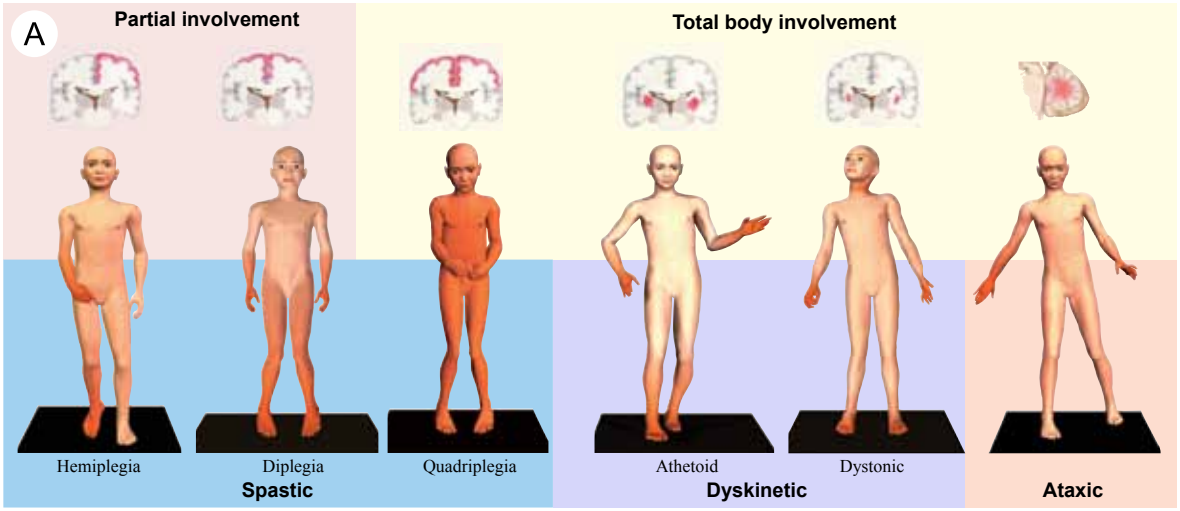
Absent traction response indicating poor head control in a 10 month old baby.



Increased tone in the limbs and truncal hypotonia is common in spastic quadriplegia.

H **Major deficits in patients with CP**

- Loss of selective motor control and dependence on primitive reflex patterns for movement
- Abnormal muscle tone that is strongly influenced by body posture, position & movement
- Imbalance between agonist and antagonist muscles that, with time and growth, leads to fixed muscle contracture and bony deformity
- Impaired body balance mechanisms
- Sensory loss
  - Vision
  - Hearing
  - Superficial & deep sensation
- Associated problems
  - Seizures
  - Mental retardation
  - Behavior problems
  - Nutrition
  - Constipation

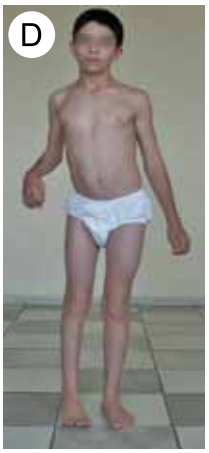


Clinical classification B

Tonus	*Lesion site*
Spastic	Cortex
Dyskinetic	Basal ganglia - extrapyramidal system
Hypotonic / Ataxic	Cerebellum
Mixed	Diffuse

Anatomical classification C

Location	*Description*
Hemiplegia	Upper and lower extremity on one side of body
Diplegia	Four extremities, legs more affected than the arms
Quadriplegia	Four extremities plus the trunk, neck and face
Triplegia	Both lower extremities and one upper extremity
Monoplegia	One extremity (rare)
Double hemiplegia	Four extremities, arms more affected than the legs



All hemiplegic children become independent walkers by the age of 3. Sensory deficits and learning disability add to the movement problem in hemiplegia. Prognosis for independent living is good.

Most diplegic children need various treatments to be able to walk independently. Problems in maintaining balance and spasticity interfere with walking. Children who can sit by the age of 2 can usually walk by the age of 4 to 7. Hand dexterity is impaired. Children have difficulty writing or other tasks that need fine motor control. Almost all diplegic children need surgery for contractures and deformities, many use walking aids.

Classification

CP encompasses a spectrum of motor disorders of varying tone, anatomical distribution and severity [A]. Clinicians classify patients to describe the specific problem, to predict prognosis and to guide treatment. Classification is based on the change in muscle tone [B], anatomical region of involvement [C] and severity of the problem. Classification provides a clearer understanding of the specific patient and directs management.

The predominant types of motor impairment are spastic, dyskinetic (dystonia and choreoathetosis) and ataxic. The spastic type can be further classified according to the distribution as hemiplegia, diplegia and quadriplegia. Even though they are clinically imprecise and may lack reliability among observers, these terms are conceptually useful.

Spastic CP

Spasticity is defined as an increase in the physiological resistance of muscle to passive motion. It is part of the upper motor neuron syndrome characterized by hyperreflexia, clonus, extensor plantar responses and primitive reflexes. Spastic CP is the most common form of CP. Approximately 70% to 80% of children with CP are spastic. Spastic CP is anatomically distributed into three types.

Hemiplegia

With hemiplegia, one side of the body is involved with the upper extremity generally more affected than the lower [D]. Seizure disorders, visual field deficits, astereognosis, and proprioceptive loss are likely. Twenty percent of children with spastic CP have hemiplegia. A focal traumatic, vascular, or infectious lesion is the cause in many cases. A unilateral brain infarct with posthemorrhagic porencephaly can be seen on magnetic resonance imaging (MRI).

Diplegia

With diplegia, the lower extremities are severely involved and the arms are mildly involved [E,F]. Intelligence usually is normal, and epilepsy is less common. Fifty per cent of children with spastic CP have diplegia. A history of prematurity is usual. Diplegia is becoming more common as more low- birth-weight babies survive. MRI reveals mild periventricular leukomalacia (PVL).

Quadriplegia (Total body involvement - tetraplegia)
With quadriplegia, all four limbs, the trunk and muscles that control the mouth, tongue, and pharynx are involved [A and B]. When one upper extremity is less involved, the term triplegia is used. Thirty percent of children with spastic CP have quadriplegia. More serious involvement of lower extremities is common in premature babies. Some have perinatal hypoxic ischemic encephalopathy. MRI reveals PVL.

Dyskinetic CP
Abnormal movements that occur when the patient initiates movement are termed dyskinesias [C,D]. Dysarthria, dysphagia, and drooling accompany the movement problem. Mental status is generally normal, however severe dysarthria makes communication difficult and leads the observer to think that the child has intellectual impairment. Sensorineural hearing dysfunction also impairs communication. Dyskinetic CP accounts for approximately 10% to 15 % of all cases of CP. Hyperbilirubinemia or severe anoxia causes basal ganglia dysfunction and results in dyskinetic CP.

Ataxic CP
Ataxia is loss of balance, coordination, and fine motor control [E]. Ataxic children cannot coordinate their movements. They are hypotonic during the first 2 years of life. Muscle tone becomes normal and ataxia becomes apparent toward the age of 2 to 3 years. Children who can walk have a wide-based gait and a mild intention tremor (dysmetria). Dexterity and fine motor control is poor. Ataxia is associated with cerebellar lesions.

Mixed CP
Children with a mixed type of CP commonly have mild spasticity, dystonia, and/or athetoid movements [F]. Ataxia may be a component of the motor dysfunction in patients in this group. Ataxia and spasticity often occur together. Spastic ataxic diplegia is a common mixed type that often is associated with hydrocephalus.

Exceptions
Some children with CP cannot be fitted into these CP groups because they present with many different impairments. Dystonia may be seen in the spastic child, and anatomical classification may not be fully explanatory because clinical findings may overlap. An example is the hypotonic total-body-involved baby who stays hypotonic throughout childhood. Define the pathological abnormalities observed in these children according to the anatomical, and clinical involvement, as described above.

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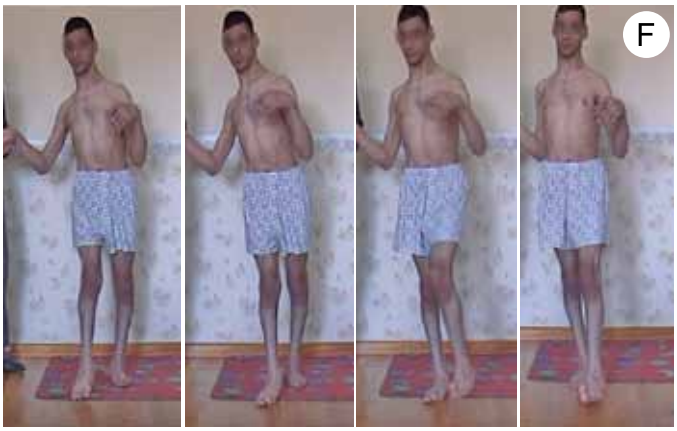
Total body involved children have various associated impairments. The severity of the motor involvement and the associated problems prevent independent living.

Dystonia is characterised by slow and twisting movements or sustained muscle contractions in the head, trunk and extremities. Children have difficulty in sitting, most cannot walk or talk. Despite the severe movement disorder, intelligence may be normal.

Ataxia is often combined with spastic diplegia. Most ataxic children can walk, but some need walkers.



The movement problem in mixed CP is a combination of spasticity, dystonia and ataxia. Involuntary movements combined with spasticity and muscle weakness may make independent mobility impossible. Upper extremity involvement prevents walker use.

A	Associated problems in CP
	Seizures
	Visual impairments
	Intellectual impairment
	Learning disabilities
	Hearing problems
	Communication problems and dysarthria
	Oromotor dysfunction
	Gastrointestinal problems and nutrition
	Teeth problems
	Respiratory dysfunction
	Bladder and bowel problems
	Social and emotional disturbances

B	Primary impairments due to the neurological lesion
	Cortical blindness
	Deafness
	Intellectual impairment
	Epilepsy
	Secondary problems - disabilities
	Strabismus due to weak eye muscles
	Malnutrition due to swallowing deficits
	Tertiary problems - handicaps
	Loss of binocular vision
	Psychosocial problems

C

Primary impairment	Secondary impairment	Deprivation handicap (tertiary impairment)
Eye muscle weakness	Strabismus	Loss of binocular vision
Muscle weakness loss of balance	Impaired mobility	Loss of stimulation & play
Loss of sensation in the hand	Neglect of the involved hand	Failure to develop hand function

Multiple seizures complicate the management of this child. They also lead to mental retardation. Children with mental retardation cannot cooperate with rehabilitation efforts.

E	Visual impairments seen in CP
Pathology	*Clinical finding*
Damage to the visual cortex	Cortical blindness
Damage to the optic nerve	Blindness
Loss of oculomotor control	Loss of binocular vision
Refraction problems	Myopia

Associated Problems

A number of associated problems occur that increase with disease severity [A]. Cortical blindness, sensory loss, deafness, mental retardation and epilepsy are primary impairments because of the neurological lesion. Disabilities that are secondary to motor deficits are weakness of external eye muscles causing strabismus or difficulties in normal swallowing leading to malnutrition. Malnutrition is an important cause of retarded brain growth and myelination. Lastly, deprivation handicaps occur [B]. The child who cannot move is deprived of peer interaction and stimulation through play. Psychosocial problems develop as a result.

Check for the presence of associated problems and get appropriate referral for treatment. Correct these problems as much and as early as possible to prevent the development of deprivation handicaps [C].

Intellectual impairment

Cognition refers to specific aspects of higher cortical function; namely, attention, memory, problem solving and language. Cognitive disturbance leads to mental retardation and learning disability. The prevalence of moderate, severe and profound mental retardation is 30 to 65% in all cases of CP. It is most common in spastic quadriplegia. Visual and hearing impairments prevent the physician from accurately assessing the degree of intellectual impairment. Children with intellectual impairment need special education and resources to stimulate the senses for optimal mental function.

Epileptic seizures

Seizures affect about 30 to 50% of patients [D]. They are most common in the total body involved and hemiplegics, in patients with mental retardation and in postnatally acquired CP. Seizures most resistant to drug therapy occur in hemiplegics. Seizure frequency increases in the preschool period. Electroencephalograms are necessary for the diagnosis of seizure disorder.

Vision problems

Approximately 40 % of all patients have some abnormality of vision or oculomotor control [E]. If there is damage to the visual cortex, the child will be functionally blind because he will be unable to interpret impulses from the retinas. In severe cases, the optic nerves may also be damaged [F]. Loss of coordination of the muscles controlling eye movements is very common [G]. The child cannot fix his gaze on an object. In half of the cases, binocular vision does not develop. Myopia is a concomitant problem. Screen for visual deficits because some are preventable and they contribute to the movement problem.

Total body involved blind child

Strabismus interferes with binocular vision in this child. Rehabilitation of visual problems may be possible in CP to a certain extent.

Hearing

Sensorineural hearing loss is seen in 10 % of children [A]. Children born prematurely are at high risk for hearing loss. It is generally not diagnosed early because of other handicaps. Test all babies for hearing loss because appropriate hearing devices prevent many future problems resulting from loss of hearing ability.

Communication problems and dysarthria

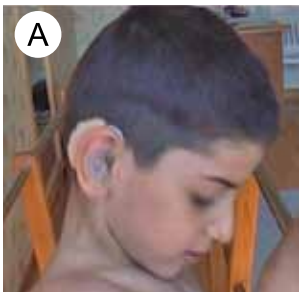
Dysarthria refers to speech problems. The child has difficulty producing sound and articulating words. Dysarthria occurs in 40% of patients. The causes are respiratory difficulties due to respiratory muscle involvement, phonation difficulties due to laryngeal involvement, and articulation difficulty due to oromotor dysfunction. Spasticity or athetosis of the muscles of the tongue, mouth and larynx cause dysarthria. It is important that every child is provided with an alternative means of communication as early as possible to avoid further disability [B,C].

Oromotor dysfunction

The child has difficulty sucking, swallowing, and chewing [D]. Drooling [E], dysarthria and inability to eat result in failure to thrive, delayed growth and nutrition, poor hygiene [F,G] and impaired socialization.

Gastrointestinal problems and nutrition

There is a general deficiency of growth and development. Children with dyskinesia and spastic quadriplegia fail to thrive [H,I]. This is related to inadequate intake of food [K], recurrent vomiting with aspiration secondary to gastroesophageal reflux and pseudobulbar palsy. Difficulties in swallowing (dysphagia), hyperactive gag reflex, spasticity or loss of fine motor control impair feeding. Gastroesophageal reflux and impaired swallowing cause aspiration pneumonia. Many children with CP have high basal metabolic rates. Increase in basal metabolic rate coupled with feeding difficulties cause malnutrition. Malnutrition may be severe enough to affect brain growth and myelination in the first 3 years of life. There is immune system suppression and increased risk of infection.



This severely involved diplegic child with hearing impairment has been using hearing aids ever since he was a baby.

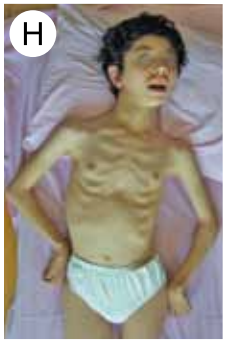
Communication aids range from advanced computer systems to simple picture boards. Children with adequate mental function learn to use these to interact with their environment.

D	Oromotor dysfunction
	Drooling
	Dysarthria
	Inability to chew
	Inability to swallow

Mouth hygiene is poor and dental caries is common. Obtain regular dental care.

G	Teeth problems	
	Dentin	Primary or hyperbilirubinemia
	Malocclusion	Spasticity
	Tooth decay	Feeding, swallowing problems
	Gingival hyperplasia	Antiepileptic drug use

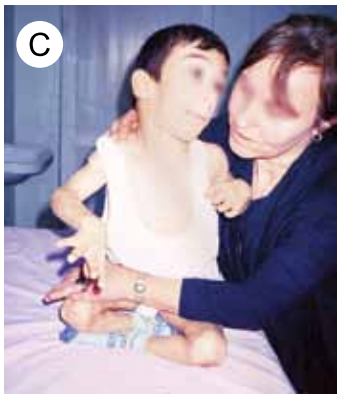
I	Reasons for failure to thrive
	Inadequate food intake
	Recurrent vomiting
	Aspiration
	High basal metabolic rate

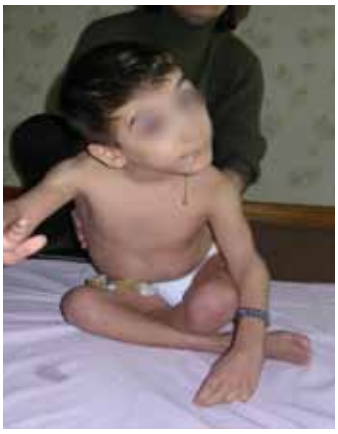


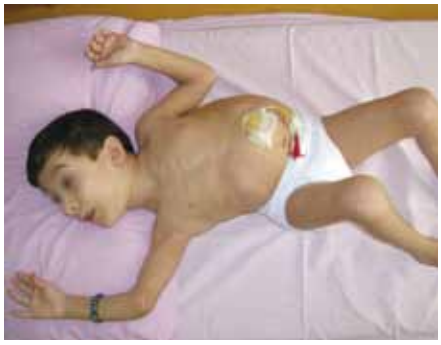
Spastic quadriplegic child with malnutrition

K	Causes of inadequate food intake
	Difficulty chewing and swallowing
	Hyperactive gag reflex
	Spasticity of oropharyngeal muscles
	Loss of selective control of oropharyngeal muscles
	Gastroesophageal reflux

Drooling and strabismus coexist in this child. Drooling is caused by oromotor dysfunction and is a difficult problem to treat. Consider oral medications and botulinum toxin in management.







The severely involved mixed quadriplegic child seen above was 14 years old and weighed only 15 kgs. He could not speak, chew or swallow. He had drooling and gastroesophageal reflux. After his esophageal ulcers bled twice, he had a gastrostomy tube inserted 2 years ago. He has been steadily gaining weight ever since.

Respiratory problems

Aspiration in small quantities leads to pneumonia in children who have difficulty swallowing. Premature babies have bronchopulmonary dysplasia. This leads to frequent upper respiratory tract infections. Respiratory muscle spasticity contributes to the pulmonary problems.

Bladder and bowel dysfunction

Loss of coordination of bowel and bladder sphincters results in constipation and/or incontinence. Enuresis, frequency, urgency, urinary tract infections and incontinence are common problems [A]. The causes are poor cognition, decreased mobility, poor communication and neurogenic dysfunction [B]. Urodynamic assessment has demonstrated bladder hyperreflexia, detrusor sphincter dyssynergia, hypertonic bladders with incomplete leakage and periodic relaxation of the distal sphincter during filling.

Constipation is a common but overlooked phenomenon. It causes distress in the child, increases spasticity and results in poor appetite. It is a result of many factors, including poor diet and decreased mobility. Establishing a routine for bowel training and encouraging upright posture help reduce constipation.

Psychosocial problems

A diagnosis of CP is extremely stressful for the family and the child when he grows up. This causes various reactions ranging from denial to anger, guilt and depression. Coping with the emotional burden of disability is easier if the family has strong relationships, financial security, and supportive members of the community. The child and the family need to find ways to connect to each other. A healthy relationship between the mother and the child forms the basis of future happiness.

Prevention or appropriate treatment of associated problems improves the quality of life of the child and the family [C].

A Urinary problems
Enuresis
Frequency
Urgency
Urinary tract infections
Incontinence

Causes of urinary problems B
Poor cognition
Decreased mobility
Decreased communication skills
Neurogenic dysfunction

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Physical Examination and Making the Diagnosis

Physical examination of a child with movement problem has two basic purposes [A]. First, physical examination accompanying a detailed history enables an accurate diagnosis. Second, it allows the treating physicians to define the impairments and disabilities, determine the functional prognosis and set treatment goals in children with CP. These then help devise a treatment plan for each child.

It is difficult to identify the cause of CP. When faced with a motor disorder in the child, the physician must be careful to rule out conditions that are results of genetic defects, such as hereditary spastic paraplegia, that are similar to CP. A detailed history and physical examination help the clinician exclude these rare syndromes and prevent expensive and extensive work-up.

Physical examination of the child with CP is not easy. It is a three-way relationship between the child, the physician and the family [B,C]. Adjustment problems can cause fear, distrust, confusion, and anxiety in the family and in the child. This disturbs their capability to understand the problem and cooperate with the treatment team. The physician must be willing to deal with anxious, confused, frustrated and unhappy families and frightened children. The examination cannot succeed unless the physician gains the parents' confidence and trust. Parents will trust a physician who takes a genuine interest in their child.

History

History is a key component in evaluating the child [D]. It provides valuable information for diagnosis. In children with a definite diagnosis, the timing of achievement of developmental milestones and the presence of associated impairments help to decide a functional prognosis. The physician gains insight into the parents' expectations and disappointments from previous treatment procedures. Knowledge of previous botulinum toxin injections, physiotherapy, surgical procedures, outcomes, complications, and psychological burden are key issues when making a treatment plan [E].

History taking provides the time and room to build a sense of understanding between the family and the physician. The goal is to make the child and the family comfortable so that the clinical examination will be accurate.

E — Key points in history

Health of parents	Development and present status of	
Hereditary factors	Head balance & control	
Siblings	Sitting	Convulsions
Pregnancy	Crawling	Emotional development
Labor and delivery	Walking	Social and recreational activities
Rh factor	Feeding	School placement
Birth weight	Dressing	Parental attitude
Condition at birth	Toilet care	Braces
Neonatal history	Speech	Medication
Age disability recognized and symptoms noted	Mental status	Previous treatment
	Hearing	Reason for referral
	Vision	
	Handedness	
	Drooling	

A — Goals of physical examination in a child with movement disorder

Establish an accurate diagnosis
Differentiate CP from progressive causes of childhood neuromotor disability
Classify the type and severity of involvement
Define the musculoskeletal impairment (spasticity, balance, weakness, contractures and deformities) and decide on ways of treatment
Evaluate associated impairments and get appropriate treatment
Determine functional prognosis
Set treatment goals
Devise a treatment plan
Evaluate the outcome of previous treatment procedures
Assess the changes that occur with treatment as well as with growth & development

A lot of information can be gained by watching the child move. The child is generally upset and frightened when he first enters the doctor's office. He must have some time to get used to his surroundings. In the meantime, the doctor should calmly talk and play with him while watching him. The room must be spacy, well-lit and friendly to calm the upset child. Play is a wonderful opportunity to establish a relationship.

Examination of the child with CP may be difficult and frustrating. Playing with the child and using simple objects as toys helps the physician to establish a better contact with the parents and the child.

D — A detailed history provides knowledge about

Risk factors
Timing of achievement of developmental milestones
The presence of associated impairments
Progression of child's capabilities
Insight into the family's resources

Try to be friends with the older child and the adolescent. Talk to them alone and if necessary perform the examination when members of the opposite sex are not present. Do not treat older children like babies.

Examination begins with observation. Do not touch the child before you watch her move.

Examine the young and frightened child in his mother's lap. Evaluate tonus abnormalities when the child is comfortable.

Guide the parents to help you during the clinical examination.

Start the examination by giving the child something to play with. A ball or a balloon will help to test upper extremity function.

Let the child sit in the mother's lap. Provide the older child with a chair of his or her own. Remember to smile at and talk to the child. Generally the parents provide the information. Older children should answer for themselves [A,B]. The parents can help fill in the details later. Including the child in the conversation builds trust. If the parents trust the physician, they will be more open-hearted when expressing their expectations and disappointments.

The child and the parents will remember less than 20% of the information provided. Provide them with a written summary of the results and the treatment plan for future reference.

Clinical Examination

Observing the child's movements is the initial and a crucial part of the examination. Observe before you touch [C]. If the child is young, apprehensive or tearful, let him or her stay on mother's lap while you watch and talk to the mother. As the child adapts to the environment, slowly place him or her on the examination table or on the floor and watch him or her move around. If the child cries a lot and does not cooperate, continue while he or she is in the mother's lap [D].

Tools required for the examination are very simple: toys, small wooden blocks, round beads or pebbles, triangular, circular and square shaped objects, a few coins, objects with different textures and a tape measure.

Perform a neurological, musculoskeletal and functional examination, although not necessarily in that order [E]. Every physician develops his or her own style and sequence of examination over the years [F,G].

G

Examination outline
Neurological examination
Skull, head circumference
Spine
Mental status
Cranial nerves
Vision - hearing - speech
Motor system
Muscle tone
Muscle power
Muscle bulk
Degree of voluntary control
Reflexes
Involuntary movements
Sensory examination
Sphincters
Developmental milestones
Musculoskeletal examination
Range of motion
Deformities, contractures
Posture
Functional examination
Sitting
Balance
Gait
Hand function

Neurological examination

Neurological evaluation of the infant and the child requires adequate knowledge of neurological developmental stages [A].

Mental status

Observe the child's orientation and interest in the surroundings. Watch for eye contact, following objects, alertness, and ability to obey simple commands.

Vision and hearing

The diagnosis of visual and hearing loss in infants can be easy. Call the child when he is not looking. Clap your hands or deliberately drop an object to make a noise behind the child and watch the response. If the child does not seem to hear, look in the child's ears for wax or signs of infection. Considering the high incidence of visual and oculomotor problems in cases of CP, all children with a definite diagnosis of neurodevelopmental delay and/or CP should undergo a detailed ophthalmological and audiological examinations during early infancy. The examinations should be repeated at yearly intervals until school age.

Muscle strength and selective motor control

Many children with CP cannot voluntarily contract or relax their muscles in isolation and therefore are unable to move their joints separately. For example, when the child attempts to extend his elbow, he involuntarily moves his whole arm. Lack of selective motor control makes it impossible to determine muscle strength using simple manual muscle testing [B]. Observe muscle strength by watching the child perform certain tasks, such as throwing or hitting a ball.

Reflexes

Evaluate the persistence of primitive reflexes and the absence of advanced postural reactions [C,D]. The presence of primitive reflexes beyond 6 months of age is a sign of poor prognosis [E].

Muscle tone and involuntary movements

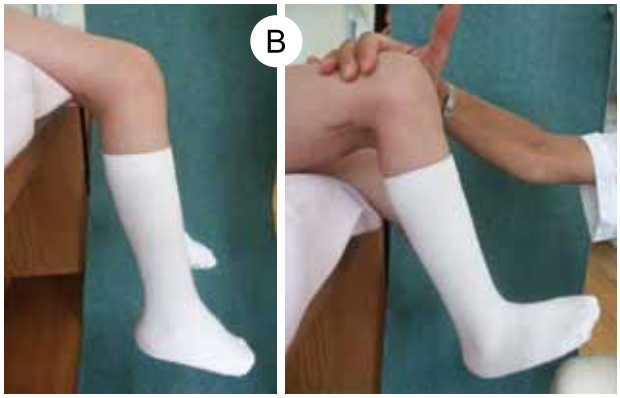
The child must be calm for assessment of muscle tone. Place the head in neutral position because turning or flexion can trigger tonic neck reflexes and interfere with muscle tone. Spasticity is the resistance felt while moving the joint through a passive range of motion. Use the modified Ashworth or Tardieu scales to grade spasticity. Also record tremor, chorea, athetosis, dystonia and ataxia [F].

This spastic quadriplegic child still has not developed protective extension and is severely delayed.

This 8 month old baby has difficulty maintaining head control on traction response indicating developmental delay.

A **Normal developmental stages of the child**

Age (months)	*Milestones*
1	Lifts head
3	Good head control, follows, laughs, smiles
5	Reaches and grasps objects
6	Propped sitting
8	Independent sitting, equilibrium reflexes
9	Gets to sitting position, presents parachute reflex
10	Pulls to stand, cruises
12-14	Walks, first words
18	Removes clothes, uses spoon
24	Uses two word phrases, throws overhand
30	Knows full name, puts on clothing
36	Jumps, pedals tricycle, learns rhymes
48	Hops, plays with others



Children who do not have selective motor control are unable to dorsiflex the ankle without moving the hip or knee; but they can produce this response in a gross flexion movement of the lower extremity. This is called pattern response. When the child flexes the hip against resistance, ankle dorsiflexion becomes apparent.

Signs of poor prognosis E

Present	*Absent*
ASTNR	Parachute response
STNR	Neck righting reactions
Moro	
Extensor thrust	
Stepping reflex	

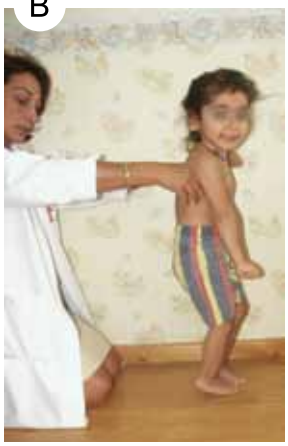
F **Differences between spasticity & dystonia**

	Spasticity	*Dystonia*
Examination	You feel	You see
Tendon reflexes	Increased	Generally normal
Clonus	Present	Absent
Pathological reflexes	Present	Rare

A

Musculoskeletal examination
Joint range of motion (ROM)
Deformities
Contractures
Balance
Posture
Sitting
Gait

B



Deformities are not apparent in many young children when they lie supine. Bring the child to erect position to demonstrate dynamic deformities.

C

Spinal deformity	Occurs in
Scoliosis	Total body involved spastic and dystonics
Kyphosis (thoracolumbar)	Patients with no sitting balance
Kyphosis (lumbar)	Patients with hamstring contractures
Hyperlordosis (lumbar)	Ambulatory patients with hip flexion contractures

D

E

Inspect the back when the child is sitting. Visual examination is sufficient to detect scoliosis or kyphosis.

Musculoskeletal examination

The musculoskeletal examination [A] reveals contractures and deformities that interfere with mobility. Perform the examination in a comfortable room with adequate space and props to attract the child's attention. Control spasticity by relaxing the child.

Range of motion

Examine range of motion in a slow and smooth manner because sudden stretch of the muscle will increase spasticity, creating the false impression of a fixed joint contracture.

Most young children do not have fixed deformities. The hip and knee joints can be moved through a full range of motion when the patient is prone or supine. However, the child will demonstrate hip flexion and adduction, knee flexion or extension and ankle equinovarus or valgus in the erect position when weightbearing. This is dynamic deformity caused by spasticity, impaired motor control and weakness of muscles [B].

Severe dynamic deformity caused by spasticity is difficult to differentiate from contracture. Stretch slowly, reassure the child and provide a relaxed and calm atmosphere in which to assess muscle tone.

Back assessment

Spinal deformity [C] associated with CP might be postural or structural and includes scoliosis, hyperkyphosis, and hyperlordosis. Patients lacking sitting balance often exhibit a long postural kyphosis. Lumbar hyperlordosis occurs in ambulatory patients with hip flexion contractures, whereas lumbar kyphosis occurs in patients with hamstring contractures. Inspect the back for scoliosis and kyphosis with the patient standing and in forward flexion. Examine the back of the nonambulatory child [D,E] while he or she sits in the wheelchair. Have the child bend forward as you check for any paramedial elevations indicating lumbar spine involvement or rib elevations showing thoracic spine involvement. Note sitting balance and pelvic obliquity, if present. Contracture and/or limb length discrepancy also contribute to spinal asymmetry.

Pelvic obliquity

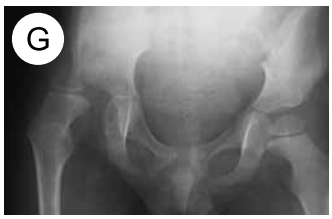
Pelvic obliquity is the abnormal inclination of the pelvis in the frontal plane. It is commonly associated with scoliosis and hip instability in the nonambulatory child. Check for sitting balance in the child with scoliosis and hip dislocation.

Limb-length discrepancy

Measure actual lower limb lengths from the anterior superior iliac spine to the medial malleolus. Measure from the trochanter to the knee joint line and from there to the medial malleolus if knee flexion contracture is present [F,G].

F

G

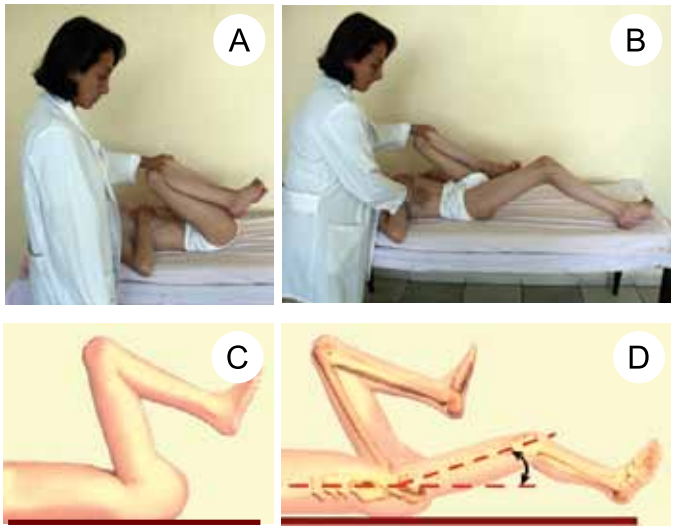


Limb length discrepancy is a sign of pelvic obliquity or hip instability. Actual limb length discrepancy may occur in hemiplegic CP due to unilateral growth asymmetry. Repeat the measurements for limb length in the upright position if you wish to prescribe shoe lifts.

Hip assessment

Measure passive and active hip range of motion. Check for flexion and adduction contractures. Evaluate flexion contracture with the Thomas test.

The Thomas test is based on the fact that a hip flexion contracture is compensated by an increase in lumbar lordosis. If the lumbar spine is fixed in the neutral position, lordosis is prevented and hip flexion contracture becomes apparent. The child lies supine on the examination table. Bring both legs up to the chest to stabilize the lumbar spine and decrease the lordosis [A]. Flex the hip and feel that the pelvis is straight. Then, keep one leg in flexion while extending the other until resistance in hip extension is felt or movement in the pelvis occurs [B]. The extended leg should be flat with the knee in full extension. If knee flexion contracture is present, extend the knee beyond the table. The angle between the thigh and the examination table gives the degree of the flexion contracture of the hip [C,D].



The Thomas test shows flexion contracture of the hip. Do not hyperflex the pelvis when flexing the hip.

Test for adduction contracture Evaluate range of abduction with the hips in flexion [E] and in extension [F]. If abduction is limited when the hips are extended but better when they are flexed, the adduction contracture is caused by gracilis and medial hamstring spasticity. If hip abduction is limited in both extension and flexion, the cause is hip adductor spasticity.

Test adduction contracture in both flexion and extension.

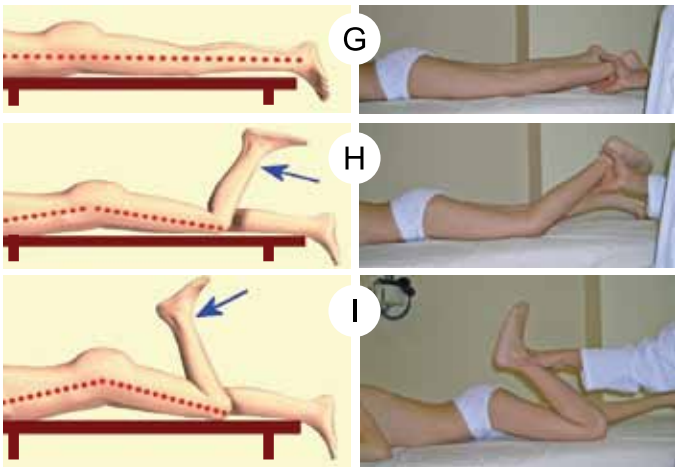
The Ely test shows rectus femoris tightness. The rectus femoris flexes the hip and extends the knee, crossing both joints so that when the hip is in extension, it is difficult to flex the knee if the rectus is tight. With the child lying prone [G], stabilize one hip in extension and bring the lower leg quickly into flexion [H]. If the buttock rises off the table, it is a sign of spastic or tight quadriceps muscle [I].

Use the Ely test to demonstrate rectus femoris spasticity and hidden flexion contracture of the hip. Most children are unhappy in the prone position so they will have increased muscle tone. Be careful not to mistake increased tone from actual contracture.

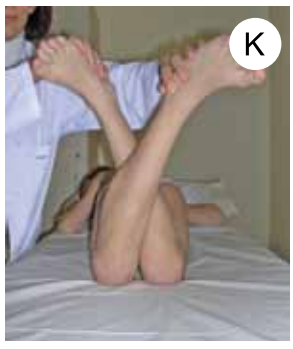
If the leg is brought into flexion swiftly, the Ely test will demonstrate rectus femoris spasticity. Do the test once more, slowly, in order to differentiate rectus femoris tightness from spasticity.

Test for hip rotation Test in prone position with the knee in flexion. Excessive internal rotation suggests persistent femoral anteversion [J,K].

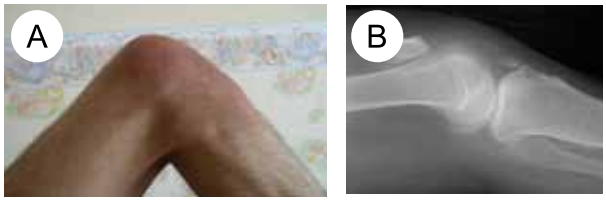
Test for hip rotation: Excessive internal rotation with the patient in prone position.



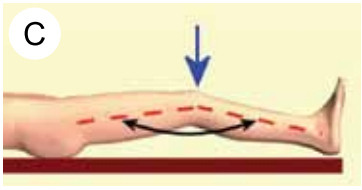
The Ely Test demonstrates rectus femoris tightness and hip flexion contracture.



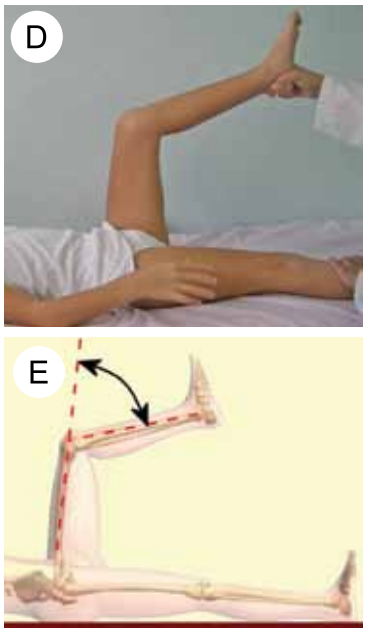
Test for hip rotation: Excessive external rotation.



Evaluation of patella position
Look for a high riding patella (patella alta) which is common in cases of rectus femoris spasticity.



Test for posterior capsule tightness
1. Extend the child's legs on the examination table.
2. Force the knees and the hips in full extension.
3. The back of the knee should touch the table.
4. Limitation indicates tight posterior capsule.



The popliteal angle
Stabilize one leg on the table, then flex the other hip to 90^{o}. Extend the lower leg until you feel resistance. Measure the angle from either the tibia and the line of full extension or the 90^{o} position to full extension. Popliteal angle shows the amount of hamstring contracture.

Knee assessment

The patella position Evaluate the patella position with the child supine and sitting. The patella slides up in children with severe quadriceps spasticity [A,B].

Posterior capsule tightness Extend the leg. If it does not extend fully, slowly force the knees and hips into full extension. Limitation indicates posterior capsule tightness [C].

Popliteal angle Measure the popliteal angle to test for hamstring contracture [D,E].

Foot and ankle assessment

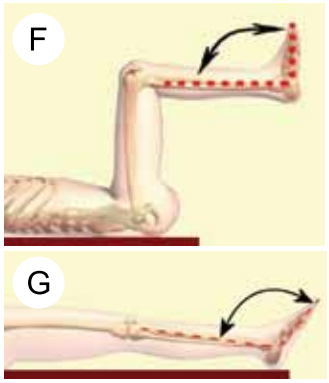
Evaluate contractures and deformities of the ankle and subtalar joints and toe deformities.

Test for triceps (gastrocnemius/soleus) contracture The gastrocnemius muscle is shortened and the soleus is normal in most children. Use the Silfverskiöld test to assess triceps surae tightness. 1. Lie the patient in supine position. 2. Measure ankle dorsiflexion first with the knee in flexion [F] and then in extension [G]. If the ankle dorsiflexion is greater when the knee is flexed, the gastrocnemius is shortened and the soleus is normal. If dorsiflexion is unchanged with the knee in flexion or extension, then both gastrocnemius and soleus are contracted. Always hold the foot in slight inversion while performing this test.

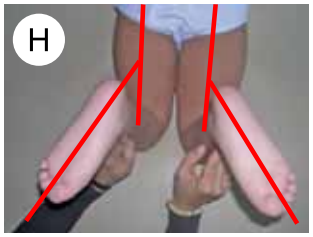
Test for tibial torsion Examine tibial torsion with the patient in the prone position. Evaluate the thigh-foot angle with the knee flexed to 90 degrees [H].

Evaluation of posterior tibialis, anterior tibialis and peroneal muscles A spastic posterior tibialis muscle causes hindfoot varus [I]. A spastic anterior tibialis muscle also causes varus and must be carefully evaluated in mono- and hemiplegic patients. A spastic peroneus or gastrocnemius muscle may cause a valgus deformity [J].

Foot deformities Pes valgus, pes varus and hallux valgus [K,L] occur in ambulatory children.

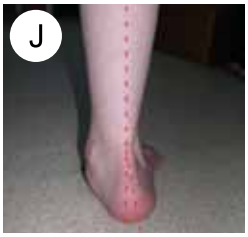


The Silfverskiold test: (The S test) shows spasticity in the triceps muscle.

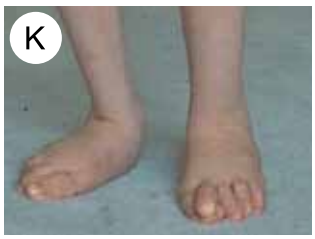


Examine external tibial torsion in the prone position with knees flexed to 90 degrees. The angle of the feet relative to the thigh (the thigh-foot angle) shows the degree of external tibial torsion.

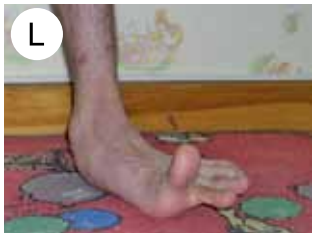
Pes equinovarus is common in hemiplegia.



Gastrocnemius spasticity is another cause of pes valgus.



Pes valgus and hallux valgus generally occur together.



Striatal toe is seen in patients with extensor hallucis longus spasticity.

Upper extremity examination

Examination for the hand and upper extremity consists of observation and evaluation of joint range of motion, the presence of contracture, muscle strength, and sensation [A]. Testing wrist and finger muscle contracture requires a detailed examination [B,C,D,E].

Spasticity of intrinsic hand muscles causes flexion contracture [F,G,H] of the metacarpophalangeal (MCP), proximal interphalangeal (PIP) and distal interphalangeal (DIP) joints. Superficial flexor tightness causes PIP joint limitation whereas deep flexor tightness causes DIP joint limitation. The most common deformity is thumb in palm deformity.

Using local anesthetic blocks to test contractures It is difficult to assess each hand muscle separately. Inject local anesthetic to the nerve to block the muscles innervated by that nerve. Then check for muscle tightness. If the muscle does not relax, fixed contracture is indicated. Blocking the spastic muscles with a local anesthetic unmasks active contraction in the antagonist muscles. Block the median nerve at the wrist to relax the wrist and finger flexors. This allows you to see activity in wrist and finger extensors. Antagonist muscle function is important when considering reconstructive surgery.

Using dynamic electromyography to test contractures Dynamic electromyography identifies which muscles are active and when they are active. Actively contracting muscles can be used for transfers. Transfers are more effective if the transferred muscle group fires in phase with the recipient muscle group.

Try to recognize adaptive responses so as not to interfere with them. Efficient hand grasp depends on balance between flexor and extensor muscles. Wrist flexors are dominant and the finger extensors are weak in the hand with spasticity. When the child wants to grasp objects, he brings the wrist into flexion by releasing his finger flexors. The child then locks the object in the palm by bringing the wrist into extension. This is not a strong grasp, but an adaptive mechanism that is valuable to the child. Flexor releases will lead to loss of hand grasp in children with this adaptive response.

Lack of sensation is a significant disability. Evaluate stereognosis, two-point discrimination, and proprioception. Stereognosis is the ability to recognize an object by touching it without looking at it. This ability requires the synthesis of multiple sensory inputs at the cortical level.

Consider using age-appropriate simple tasks to test upper extremity and hand function [I]. These include combing the hair, brushing teeth, putting food to mouth, drawing a triangle and throwing a ball.

The appearance of the hand is important especially for older girls with hemiplegia since the hand is a social tool for communication. Evaluate the appearance of the hand and the patient's satisfaction with it.

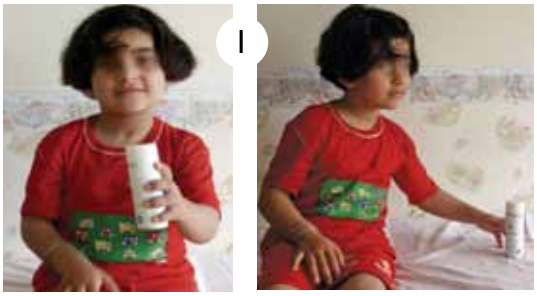
H	Flexion contracture
1. Measure flexion contracture of the wrist with the wrist in full flexion and the fingers in full extension.	
2. Slowly and gently pull the wrist into extension while keeping the finger joints in extension.	
3. The angle of wrist with the forearm is the angle of flexion contracture.	
4. Then evaluate the PIP & DIP joints separately to determine the spastic muscle group.	

A	Examination of the upper extremity
	Joint range of motion
	Presence of contracture
	Muscle strength
	Coordination
	Sensation
	Function

1. The small child sits in the mother's lap and the older in a comfortable chair.
2. A desk or a table is necessary for testing fine motor control and coordination. Children without sitting balance use their regular wheelchair with a tray placed on their lap.
3. Watch active shoulder and elbow movements while the child plays with a plastic or wooden cylindrical toy to detect movement disorders and spasticity.
4. When the child grasps the object, observe the gross motor function of the hand, the quality of grasp and release and the ability to cross the midline. For wrist and finger fine movements, small plastic beads or toys are better.

E: Courtesy of M. Carlsson

Volkmann angle demonstrates wrist flexion contracture.

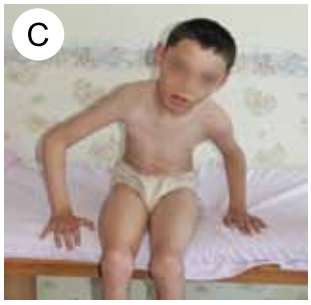


Note the speed and precision of movement and the quality of grasp and release. Observe how much the child uses his involved extremity.

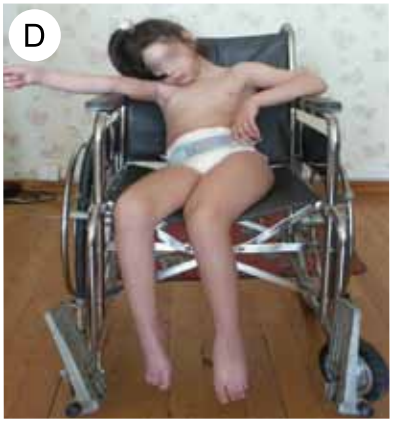
A	Classification of sitting ability
Hands-free sitter (Independent sitter)	can independently come to a sitting position, does not need hands to sit up and can sit in a normal chair without losing his balance.
Hand-dependent sitter	uses hands for support when sitting, needs a chair with side supports to be able to use his hands for eating or writing.
Propped sitter	has to be brought to a sitting position by someone else, needs external support and sits in a reclining position when strapped into the seat.

This classification applies particularly to quadriplegic children as a guide to the use of seating aids.

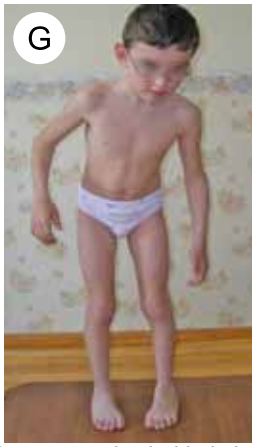
Hands-free sitter



Hand-dependent sitter



Propped sitters needs external support to sit.



The Romberg sign: the child cannot maintain his balance with eyes closed.

Functional examination

Sitting

Evaluate sitting [A] to decide whether the child needs support [B-E]. Children with adequate sitting balance are more functional.

Balance

Balance and equilibrium reactions are prerequisites for walking. Evaluate balance in all children. Push the standing child gently from the front, back, and side to see whether he or she can promptly regain balance. Assess deficiency of balance and equilibrium using the Romberg sign, unilateral standing balance test and the hop test.

Romberg sign [F,G] shows whether the child can maintain balance. If the child sways and cannot keep his balance with feet held together and eyes closed (positive Romberg's sign), then there is sensory ataxia. If the Romberg sign is negative in the ataxic child, the ataxia is of cerebellar origin.

Unilateral standing balance test [H,I] Reveals inability to maintain balance in less severely involved children. A 5 year old should be able to stand on one foot for 10 seconds. Failure in the unilateral standing balance test explains why children sometimes show excessive trunk leaning when walking.

Hop test Boys can hop on one leg for five to 10 times from age 5 years and girls from age 4 years onwards. Inability to perform single-leg hop is another sign of poor balance and neuromuscular control.

Mobility

A crucial part of the examination is the observation of the child's walking pattern [J]. Video recordings of the child's movement also guide treatment. Ask the family to obtain photographs or video recordings of their child to understand how the child functions at home. Computerized gait analysis is possible in advanced centers. The nonambulatory child is placed on the floor to assess his mobility [C-F on next page]. The child may roll, creep, crawl or 'walk on all fours'.

Unilateral standing balance test: The child is able to stand on her right foot but tends to fall when she stands on her left foot. This is a sign of unilateral balance disturbance.

J	Classification of ambulation
Community ambulators	are free to ambulate in the community independently with or without orthotics or assistive devices.
Household ambulators	walk independently indoors using braces and assistive devices. They need a wheelchair for outdoor mobility.
Therapeutic ambulators	walk as part of a therapy session for short distances with a helper. They need a wheelchair at all other times.
Nonambulators	use the wheelchair for mobility.

A

Functional scales used in CP		
Scale	*Ages*	*Measures*
Gross Motor Function Measure	Birth to 5 years	Change in gross motor function over time compared to normal children
The Pediatric Evaluation of Disability Inventory	6 months to 7 years	Functional status and functional change
Wee Functional Independence Measure	6 months to 7 years	Level of independence in 6 different areas
The Movement Assessment of Infants	Birth to 12 months	Gross and fine motor performance of infants

Functional scales

Different scales are used to assess the functional status of patients with CP [A]. Some are descriptive and compare the child with normal age-matched peers whereas a few of them measure change over time that occurs with growth and treatment. Functional tests identify babies and children who have delayed gross or fine motor development and record the progress of those children under treatment. Quality of life is measured with scales such as the CP-QOL or the CPCHILD scale.

Study, skill and experience render the application and scoring of most of these scales easier. Instructional courses are required to apply the Wee Functional Independence Measure (WeeFIM) and the Pediatric Evaluation of Disability Inventory (PEDI). These scales are expensive and many take at least an hour to administer which makes it difficult to use in outpatient clinics. The busy clinician must decide on his or her methods of assessing the functional status of the child and the effect of treatment. The Gross Motor Function Measure (GMFM) and the Gross Motor Classification System (GMFCS) have been the most helpful to date and they are freely available for use.

Gross Motor Function Measure (GMFM)

The GMFM was developed to measure changes in gross motor function over time in children with CP. It compares the child with normal children of the same age. The GMFM is a reliable scale to evaluate gross motor function. It measures the child's skill in lying, rolling, sitting, crawling, kneeling, standing, walking, running, and jumping, but it does not measure the quality of movement. It can be used for children from birth to 5 years of age.

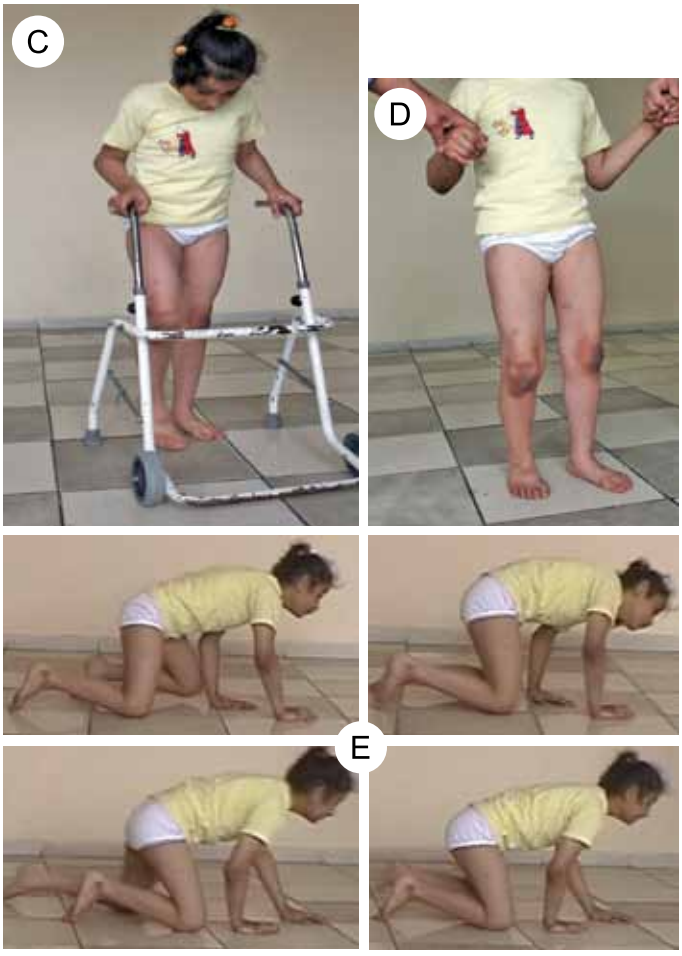
Gross Motor Function Classification System (GMFCS)

The Gross Motor Function Classification System (GMFCS) [B] was developed to create a systematic way to describe the functional abilities and limitations in motor function of children with CP. The emphasis is on sitting and walking. The purpose is to classify a child's present gross motor function. Five levels are used in the GMFCS from very mild to very severe. The levels are based on the functional limitations, the need for assistive technology and wheeled mobility. The quality of movement is not very important. Because motor function depends on age, separate scales are used for different age bands. Classification at 2 years allows one to predict prognosis at age 20 years.

The GMFCS is an important tool for physicians and therapists treating children with CP. It is easy to use; classifying a child takes 5 - 15 minutes. Physicians and therapists from various

B

Gross Motor Function Classification System (GMFCS)	
Level	*Ability*
1	Walks without restrictions
2	Walks without assistive devices but limitations in community
3	Walks with assistive devices
4	Transported or uses powered mobility
5	Severely limited dependent on wheelchair



This ten year old girl with mixed CP can use a walker for ambulation. A closer look at her knees shows callosities on both patellae. This indicates that her primary means of mobility is crawling on all fours.

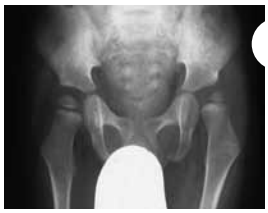
Reciprocal movement is the ability to move one extremity after the other while crawling or walking. It is a sign of good motor control.

A Progressive disorders resembling CP
Glutaric aciduria Type I
Arginase deficiency
Sjögren – Larsson syndrome
Metachromatic leukodystrophy
Lesch - Nyhan syndrome
Joubert syndrome
Chiari Type I malformation
Dandy - Walker syndrome
Angelman syndrome
Gillespie syndrome
Marinesco - Sjögren syndrome
Ataxia - Telangiectasia
Hexoaminidase A and B deficiency
Behr syndrome
Serotendinosus xanthomatosis

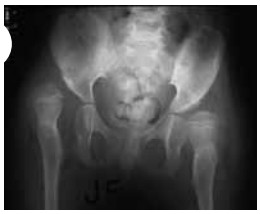
Nonprogressive disorders resembling CP B
Mental retardation
Deprivation
Malnutrition
Non-motor handicaps (blindness)
Motor handicaps (spina bifida, myopathies)

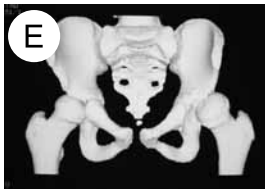
Early differential diagnosis in developmental disability

C	*Cerebral palsy*	*Mental retardation*
Risk factors	Often positive	Mostly absent
Complaints	Irritable, sleepless baby	Easy baby
Milestones	Delayed	Delayed
Examination	Delayed growth or negative	Negative or a syndrome
Muscle tone	Increased	Hypotonia
Primitive reflexes	Persist	Normal disappearance
Postural reflexes	Delayed appearance	Delayed appearance
Focal signs	Appear	Absent



D





E

Radiographies are the standard evaluation method for hip instability. A three dimensional CT scan provides valuable data in the preoperative evaluation for hip reconstruction.

disciplines can easily use this scale for their patients. Therefore, it provides a basic understanding of the level of involvement of a child for all those involved in caring for the child. The use of the GMFCS is becoming increasingly common in CP clinics as a universal tool for communication with colleagues, determining the prognosis and planning treatment.

Manual Ability Classification System (MACS)

Hand use is crucial for children's independence in daily life, to be able to attend the school and occupy themselves during leisure. The development of hand function does not follow that for gross motor development, thus there is a need to highlight the importance of hand function for independence in daily life. The Manual Ability Classification System has been developed to classify how children with cerebral palsy use their hands when handling objects in daily activities. The classification is designed to reflect the child's typical manual performance, not the child's maximal capacity. It classifies what children do when using one or both of their hands for activities, rather than assessing and classifying each hand separately.

MACS can be used for children of different ages (4-18 years), but the interpretation of the levels needs to be related to the age of the child.

Quality of Life Scales

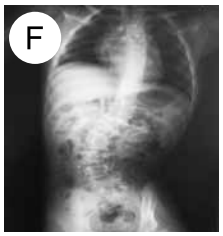
Qualtiy of life refers to an individual's subjective experience of their life and their wellbeing. QOL measures are concerned with how people feel about their lives. CP QOL - Child was first designed to assess the QOL of children with cerebral palsy aged 4-12 years. It is an age and condition specific measure of physical, mental and social wellbeing. The CPCHILD© is a measure of caregivers' perspectives on the health status, comfort, well being, and ease of caregiving of children with severe developmental disabilities, and is useful to determine the health related quality of life of these children. Some scales and suggestions for further reading are provided in the appendix.

Differential diagnosis

One needs to distinguish CP from progressive disorders of childhood [A]. It may not be always necessary to find the exact cause because this does not change the management for most children (with the exception of inborn errors of metabolism that can be cured). Mental retardation syndromes, attention deficit disorder, autism and non-motor handicaps such as blindness and emotional disorders also cause motor delay [B,C]. Cognitive problems are prominent in all these syndromes except for blindness. On the contrary, motor problems are predominant in CP. All children with suspected motor delay should be seen by a pediatric neurologist to assess for differential diagnoses.

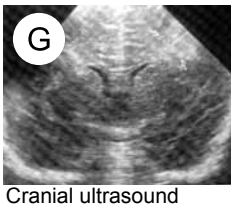
Imaging studies

Imaging studies enable the physician to define the type and location of the brain lesion and to differentiate progressive neurological syndromes.



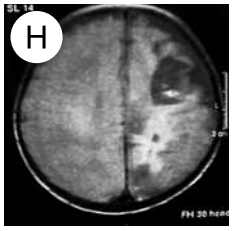
F

Evaluate scoliosis with radiographies



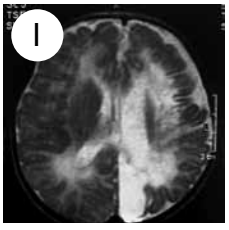
G

Cranial ultrasound



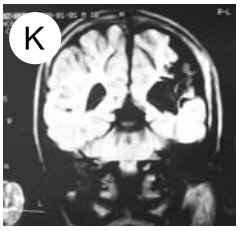
H

Porencephaly on cranial CT



I

Periventricular leukomalacia on MRI



K

Porencephaly on MRI

Radiology

The primary indication for a radiography is monitoring hip instability. Obtain baseline spine and hip radiographs in every child and follow the hip at risk with hip radiographs [D on previous page]. Measure the Reimer's index which is the percentage of femoral head coverage by the acetabulum. Three-dimensional CT is useful when planning hip reconstruction [E on previous page]. Clinical examination is sufficient to diagnose and follow-up scoliosis. Measure the Cobb angle in children who are candidates for surgery [F on previous page]. Obtain radiographs of the extremities for patients if you plan osteotomies. Standing radiographs of the feet help if there are varus/valgus deformities.

Cranial ultrasonography (USG)

Cranial USG [G on previous page] is sueful in the differential diagnosis of the infant when the fontanelle is open. It is easy and it does not require sedation as does MRI. Cranial USG evaluates the ventricles, basal ganglia and corpus callosum for eriventricular white matter ischemic injury and intraventricular haemorrhage.

Cerebral computerized tomography (CT)

CT is helpful in the diagnosis of intracranial bleeding in the newborn, it may show congenital malformations and PVL [H on previous page] but MRI is definitely superior in such lesions.

Cranial magnetic resonance imaging (MRI)

MRI is the best method for diagnosing lesions in the white matter after 2 to 3 weeks of age [I, K on previous page]. At present, MRI and ultrasonography are the only methods to show periventricular leukomalacia in an infant from one week of age. No biochemical methods are available to identify high-risk infants at birth.

Electroencephalography (EEG)

EEG measures electrical activity on the surface of the brain. It is a necessary tool in the diagnosis and follow-up of seizures.

Explanation of the diagnosis to parents

The diagnosis takes time. The child must be at least a year old before a definite diagnosis can be made, especially in cases in which the lesion occurs during the prenatal or perinatal stage (approximately 80% of all CP cases). The infant with a cerebral dysfunction shows signs of neurodevelopmental delay on initial examination. Tone abnormalities such as spasticity and dystonia, or disorders of movement such as ataxia appear generally at or after 12 months of age, when it will be possible to name the movement disorder as CP. Wait until then for a definite diagnosis while providing those babies who show delayed development with an adequate exercise program to stimulate the CNS.

The diagnosis of CP labels the child as handicapped or abnormal. Be cautious and avoid initial use of the term *cerebral palsy* to the parents because of the stigma. Instead, describe the child's symptoms and why they occur. Parents are anxious to learn about their child's problems and need definite answers. Evaluate the child's condition and prognosis well in order to provide satisfactory answers without raising false hopes or breaking all hope [A].

The diagnosis is difficult in the infant. The extent and severity of involvement that a child will have in the future is impossible to assess when he is a baby. Do not talk about the child's problems in detail. This causes disappointment in the parents who start to feel bad about their child. Slowly prepare parents for the possibility of damage to the brain. Let the child and the family bond together early in infancy. Be very cautious when predicting the future because the outcome is variable.

How to explain the diagnosis to the parents?

Your baby has had an injury to his brain. He is developing a little slower than his peers. He may have problems with movement. The outlook depends upon how serious or how extensive the brain damage is. Because he is a baby, right now it is difficult to tell what the future will be. We have to follow him carefully, he has to have therapy and we will see how he will develop. All children develop along a certain pathway. Children who have this brain injury that we call cerebral palsy also develop but they do it more slowly. If a normal child sits at the age of 6 months, he may not sit to a year and a half; if a normal child walks at one year, he may not walk until he is 3 or 4 years old. We will see how he will improve.

Your child has difficulty controlling his movements. This is because of an injury in the movement center in the brain. The brain injury is permanent. It does not get worse, but it never really gets better. Regardless, the child deserves anything we can do for him to achieve whatever potential he has. The most important things are talking, being able to eat and to take care of himself, then mobility and walking. We can help the child move better by relaxing his muscles, teaching him exercises and giving him various braces. He may need injections or surgery in the future to relieve the stiffness. Our goal is to make sure that he becomes part of the society. He must be able to communicate, take care of himself and go from one place to another. Even if his walking may be impaired he can still have a full life.

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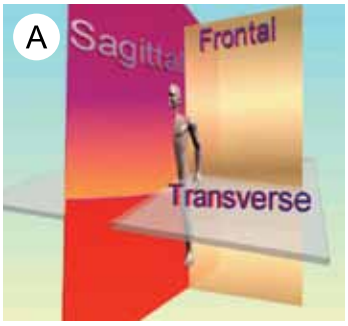
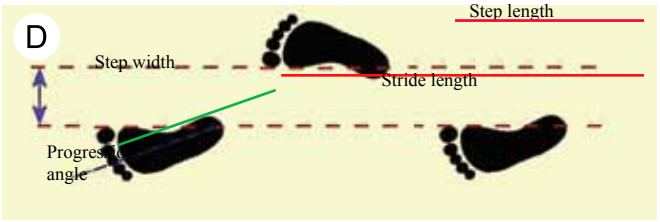


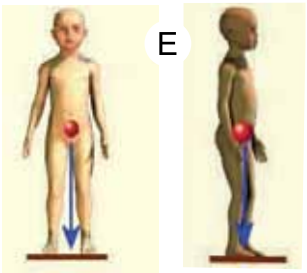
Three planes of human motion:
Sagittal: flexion / extension
Frontal: abduction / adduction
Transverse: rotation

Double support | Right foot stance, Left foot swing | Double support | Left foot stance, Right foot swing

C	Phases of gait
Stance phase	60%, foot on the ground
Swing phase	40%, foot in the air
Period of double support	11%, both feet are on the ground
Period of single support	80%, one foot is on the ground



Step length: distance from the point of contact of one foot to the point of contact of the other foot.
Stride length: distance from the initial contact of one foot to the initial contact of the same foot.
Cadence: number of steps taken per minute.



Center of mass and center of pressure
The center of mass (COM) is in front of the second sacral vertebra in a standing adult. A vector perpendicular to the ground drawn from this center of mass represents the force exerted by the body on the ground. This vector must coincide with the center of pressure (COP) of the body. The COP is a point on the ground between the feet. When the COM is over the COP, the person is balanced. When the COM is outside the COP, the person loses balance.

The body weight exerts a force to the ground. The ground responds by a force of equal magnitude and in opposite direction to the body. This ground reaction force creates rotatory forces about the joints called external moments. Muscles contract and produce internal moments to keep the body stable.

Gait

Some children with CP cannot walk. Others have walking difficulty. This is generally the basic reason for seeking medical advice in CP and probably one of the most difficult to affect. To understand the gait pathology associated with CP, first understand normal gait. Walking is one of the most important functions of the human musculoskeletal system. Efficient walking requires complete coordination of the brain, spinal cord, peripheral nerves, muscles, bones and joints.

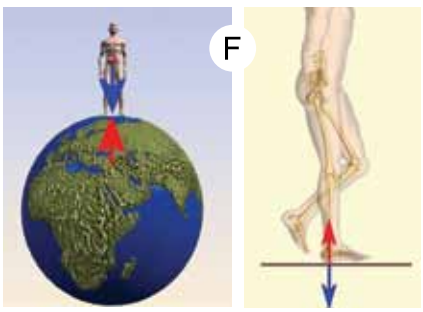
Normal gait

Walking consists of a series of movements that are repeated in a cyclical pattern. These movements are analyzed in three planes [A]. The gait cycle is defined as the period of time from the moment when one foot strikes the ground to the moment when the same foot strikes the ground again [B]. The gait cycle is divided into the stance and swing phases. The stance phase consists of 60% of the total cycle and represents the time period when one foot is in contact with the ground. The swing phase consists of 40% of the cycle and represents the period when the foot is not touching the ground [C]. In the gait cycle, an 11% period occurs during which both feet are in contact with the ground. This is the double support phase. Every person has a comfortable walking speed according to age and gender. In a healthy adult male this is approximately 80 meters per minute [D]. Normal walking requires balance, propulsion, shock absorption and energy consumption. The person must maintain balance, propel the body forward, absorb the shock created by the body weight and spend as little energy as possible.

Balance

Balance depends on the relationship between the center of mass and the center of gravity [E]. The person has balance in stance, loses it during swing and recovers it during double support. Ligaments provide static stability and muscles provide dynamic stability. Inability to maintain upright balance is an important cause of walking difficulty. During walking a person must be able to shift and support the body weight over the extremity in stance, preserving lateral balance. Children with CP lack this ability and cannot shift their weight over the leg on the ground during stance. They have a tendency to fall toward the swinging limb. This deficiency in maintaining lateral balance is the reason many diplegics and some hemiplegics walk with a sideways lurch. Weak abductors cause a sideways lurch too. Provide the patient with crutches, canes and walkers to help maintain balance.

Inability to maintain balance also creates a tendency to walk very fast, as if running. This phenomenon resembles an inexperienced rider on a bike. Inexperienced riders go fast to maintain their balance whereas experts can ride slowly without falling.



Propulsion

The body moves forward during swing and stance phases. Hip and knee flexion are necessary to clear the swinging leg off the ground. The knee must extend at the end of swing for an adequate step length. The body moves forward also over the stance leg by a series of movements called rockers [A-D]. Forward progression is disturbed in CP. The muscles cannot produce the necessary force for moving the body forward. The swinging leg cannot clear the ground because of inadequate hip and knee flexion. Step length is short because of limited knee extension. The body weight cannot move over the stance leg because of muscle weakness and contractures that disturb the rocker mechanism.

Muscle function during gait

The body mass exerts a force to the ground and the ground responds with an equal force in the opposite direction to the body; this is termed the ground reaction force. The body responds by muscle contraction to sustain balance and stability in the joints [F on previous page].

Tibialis anterior [E] is active in the first rocker of gait cycle. It allows smooth ankle plantar flexion as the foot comes in contact with the ground. It provides mediolateral stability and foot clearance by active dorsiflexion of the ankle during the swing phase. Weakness contributes to foot drag during swing and to instability during stance.

Quadriceps [F] contracts from initial contact through midstance to allow 15° of knee flexion and contribute to forward progression of body. It contracts at the end of stance to counteract the external flexor moment that the ground reaction force produces at the knee. This is a brief contraction to prevent the swinging knee from flexing too far. Weakness of the quadriceps muscle causes the knee to flex too much during stance, leading to crouch. Spasticity causes inability to flex the knee during swing leading to stiff knee gait.

Hamstrings [G] contract at initial contact to keep the hip and knee stable and at the end of swing to prevent the tibia from going too far into extension. Spasticity causes crouch.

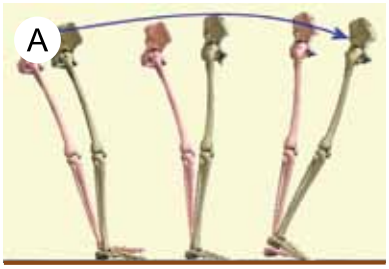
Gastrocnemius-soleus [H] are active during the middle and end of stance, limiting passive ankle dorsiflexion and providing push off. Their weakness causes crouch and spasticity causes equinus.

Shock absorption

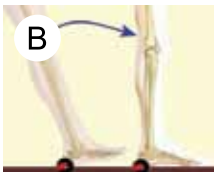
Approximately 60% of the body weight is transferred to the extremity in stance in 0.02 seconds during heel strike. The effects of this shock are reduced by muscle action at the ankle, knee and the hip. Ankle dorsiflexors limit ankle plantar flexion and allow a smooth contact with the ground. The quadriceps limits knee flexion and the hip abductors prevent excessive pelvic drop.

Energy consumption

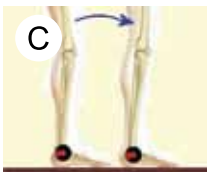
The excursion of the body center of mass [I,K] determines the energy cost of walking. Energy cost is high in patients with CP because of the increased excursion of the body center of mass.



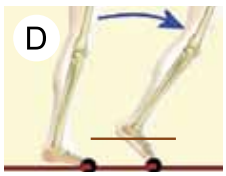
A walking person moves his body forward over the stance leg by a series of movements called rockers.



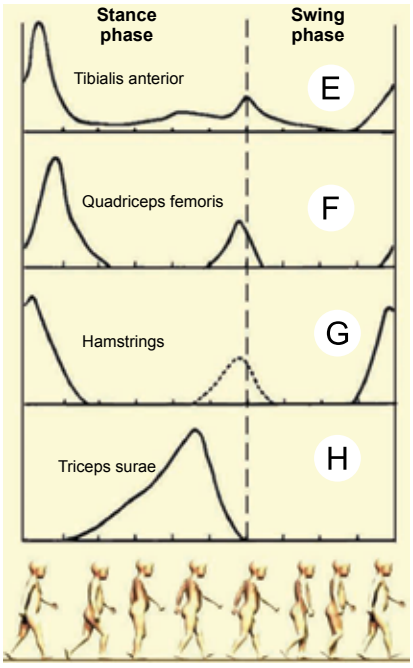
Heel (first) rocker: The period from the moment the heel strikes the ground to the moment the forefoot touches the ground. The pivot of motion is the heel.



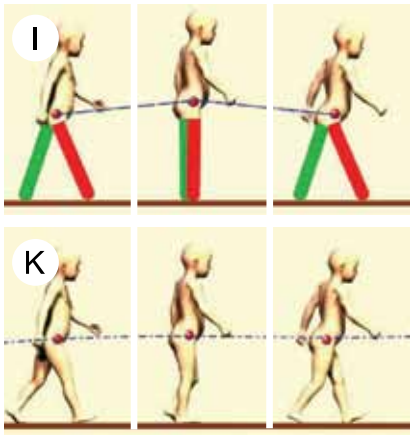
Ankle (second) rocker: The period from the moment the forefoot touches the ground to midstance. The pivot of motion is the ankle joint.



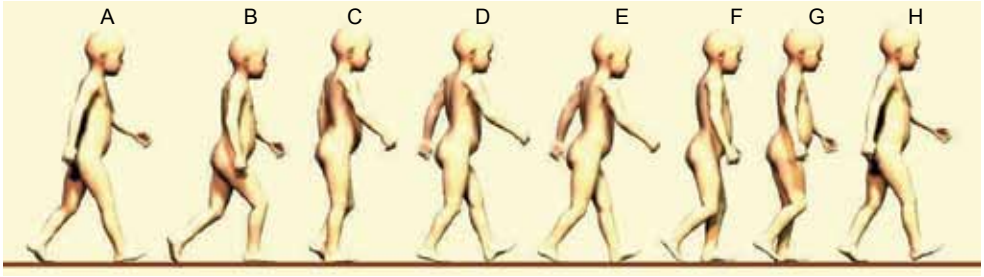
Forefoot (third) rocker: begins with heel rise and finishes with toe off. The pivot of motion is the metatarsophalangeal joint.



Timing of muscle action during gait



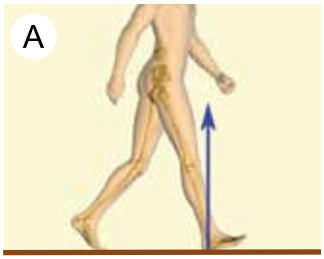
The excursion of the body center of mass is a sinusoidal curve of little amplitude. When the lower extremity joints do not move normally, the excursion increases.



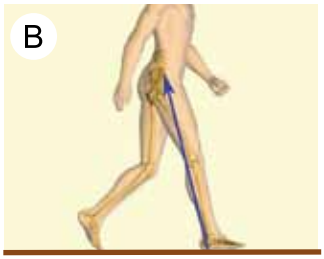
Gait cycle			
Stance phase		*Swing phase*	
A	Initial contact	F	Initial swing
B	Loading response	G	Midswing
C	Midstance	H	Terminal swing
D	Terminal stance		
E	Preswing		

Phases of Gait

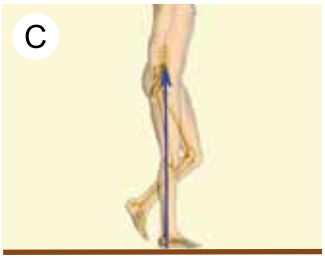
The gait cycle is divided into the stance and swing phases. Common problems in stance are positioning the foot during initial contact, too much knee, hip and ankle flexion or knee hyperextension in midstance and poor push-off. Problems in swing include insufficient foot clearance and limb advancement. Loss of stability in stance and inadequate progression in swing occur.



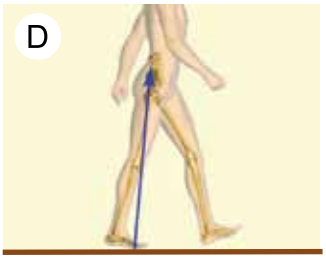
Initial contact
Initial contact begins when the foot strikes the ground. The hip is in 30º flexion, the knee in full extension, the ankle is in neutral and the foot is in supination. At the point of heel strike, the ground reaction force is in front of the hip and the knee. This creates a flexion moment at the hip and an extension moment at the knee. Hip extensors and knee flexors contract to overcome this moment. Tibialis anterior contracts to keep the ankle in dorsiflexion.



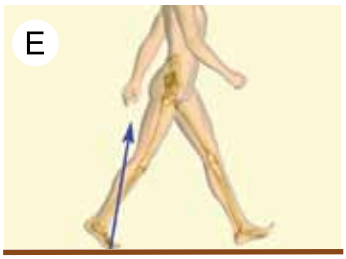
Loading response
The person puts his weight on the extremity. The hip begins to extend, the knee is in 20º and the ankle is in 10º plantar flexion. Ground reaction force creates flexion at the hip and knee, plantar flexion at the ankle. Hip and knee extensors and ankle dorsiflexors contract to counteract this force.



Midstance
Only one foot is in contact with the ground. The hip and knee are in extension, the ankle is in dorsiflexion. Ground reaction force is behind the knee and in front of the ankle, causing flexion at the knee and dorsiflexion at the ankle. Hip abductors, quadriceps and ankle plantar flexors contract.



Terminal stance
The heel begins to lift off the ground. The hip is in 10º extension, the knee starts to flex, the ankle is in plantar flexion to clear the leg off the ground. Ground reaction force is behind the hip but in front of the knee and ankle. Iliopsoas and triceps surae are active.



Preswing
The toes lift off the ground. Hip extension, knee flexion and ankle dorsiflexion increase. Ground reaction force is behind the knee producing a flexion moment. Iliopsoas, rectus femoris, triceps surae are active. Rectus femoris contracts to overcome knee flexion.

Initial swing
There is flexion at the hip and knee joints and dorsiflexion at the ankle. Hip flexors and ankle dorsiflexors are active, knee flexion is passive.

Midswing
The swing leg passes in front of the stance leg. Flexion of the lower extremity increases to a maximum. Hip and knee flexion is passive due to inertia while ankle dorsiflexors are still active.

Terminal swing
The swing leg prepares for landing. The hip is in flexion, the knee in extension and the ankle is in neutral position. Hamstring muscles contract to limit hip flexion and knee extension. Ankle dorsiflexors contract to keep the ankle at neutral.

Clinical examination of gait

Ambulatory children with CP have various types of pathological gait. Efficient intervention depends on proper evaluation [A] Observation [B] and video recordings [C] are sufficient to understand the abnormality in many cases.

Watch the video in slow motion for a better understanding. Videos are useful to demonstrate the child's progress to the parents. Computerized gait analysis is necessary in the few cases with more complex gait patterns.

Gait analysis

Computerized gait analysis is an objective, standardized, reproducible and quantifiable method to evaluate gait pathology. Computerized gait analysis consists of 5 components [D - I]. Gait analysis helps to decide on the type of therapeutic intervention and to asses the effects of the intervention. It has a role in research, education and therapeutic decision making. Computerized gait analysis has advantages and disadvantages [J].

Although gait analysis has been shown to alter decision making, there is little evidence that the decisions based on gait analysis lead to better outcomes. Gait analysis is useful as a research and education tool. It is an additional aid in decision making for treatment. It requires expensive high technology equipment and educated staff. It shows how the child walks graphically but does not tell how functional the gait pattern is unless it measures the amount of energy consumed during walking. It adds little to the clinical examination and

Examination of gait (A)
Observation
Video recording
Computerized gait analysis

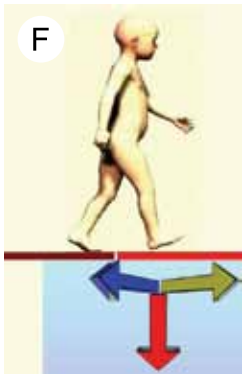
Observation (B)
1. The child walks a distance of 10 meters.
2. Stand at a distance of 3 m., watch the child walk toward you.
3. Stand at a distance of 3 m., watch from the side.
4. Look at each joint separately in the order of: L hip, R hip, L knee, R knee, L ankle, R ankle.
5. Watch balance as the child turns.
6. Record step length, stride width and any deformities.
7. Record the gait on video.
8. Do not overtire the child.

Components of computerized gait analysis (D)
Kinematics: Recording joint motion with markers and cameras
Dynamic electromyography: Electrophysiological monitoring of muscle activity using fine wire electrodes placed into the key muscles
Kinetics: Assessment of force vectors using force plates
Energetics: Evaluation of O_2 consumption and energy cost of walking using gas analysis systems
Dynamic pedobarography: Pressure changes of the sole of the feet

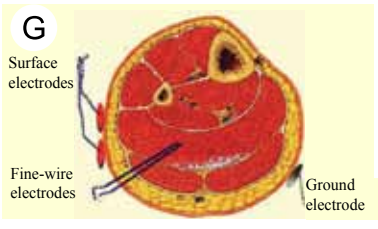
Computerized gait analysis (J)	
Advantages	*Disadvantages*
Provides quantifiable data	Data interpretation necessary
Shows moments & powers across the joints	Different laboratories produce different results for the same patient.
Shows muscle activity during gait	Expensive to start and maintain
	Difficult in small children
	Kinetic data not possible below age 4

Kinematic data are obtained from markers placed on the body. As light reflects from the marker, the camera captures it and feeds the information to the computer. Special software processes the data into graphics.

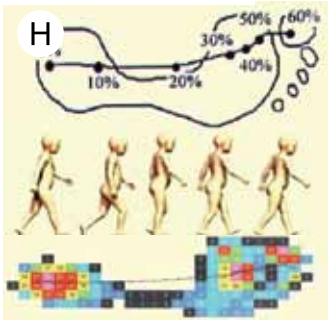
Photos E,I: Courtesy of MotionAnalysis Co.



Forces acting upon the joints are estimated through a complex mathematical equation by first capturing the ground reaction force using force plates. The three dimensions of the ground reaction force vector can be measured separately.



Needle electrodes can be placed into deep muscles and skin electrodes are used for superficial muscles in dynamic EMG.

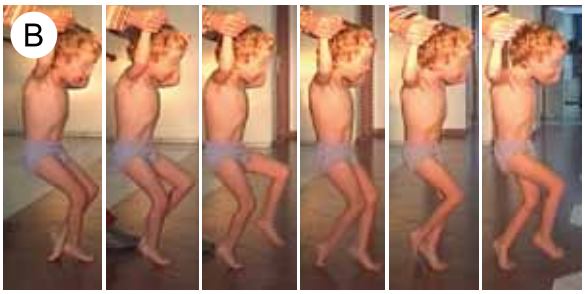


Pedobarograph measures the pressure changes of very small sections of the sole of the foot. It gives a pressure distribution map of the weight bearing foot.

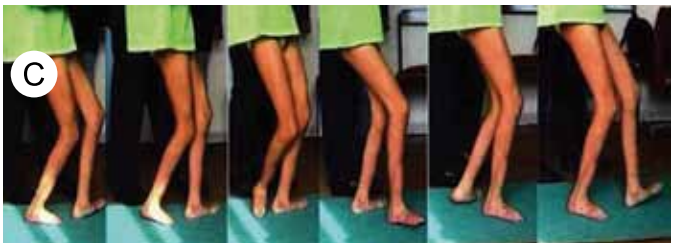
These measures are then fed into a computer system and processed. Analysis of this data by physicians experienced in the field of gait analysis will result in a clearer definition of problems during gait.

Characteristics of gait in children A

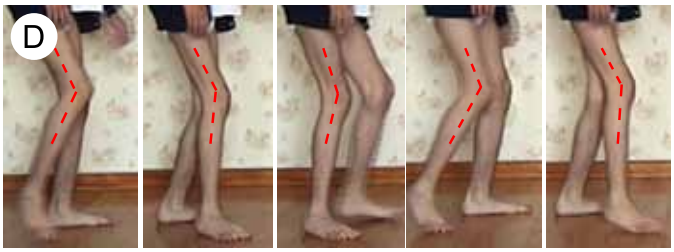
Parameter	*Characteristic*	*Normalizes at age*
Step length	Short	15
Step width	Increased	4
Cadence	Increased	15
Speed	Slow	15
Stance	Longer	4
Muscle activity	Increased	4
Heel strike	None	2-3
Knee flexion	Minimal in stance	2-3
Legs	External rotation during swing	2-3
Arm swing	Absent	4



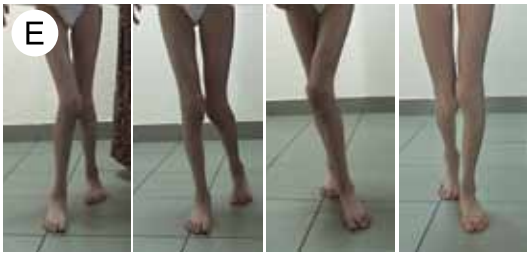
Jump gait typical for the young diplegic child.



Crouch gait occurs in the growing diplegic child. It is characterized by increased knee flexion and ankle dorsiflexion during stance. Isolated gastrocnemius lengthening or overlengthening weakens push-off and causes crouch. Severe hamstring weakness also causes crouch.



Stiff knee gait may accompany crouch. In this case, the quadriceps and the hamstring muscles are spastic. Stiff knee gait is easily recognized by shoewear due to drag in swing.



Scissoring or crossing over is caused by medial hamstring and adductor muscle spasticity in the young child. Increased femoral anteversion contributes to the problem in the older.

remains more a research tool than part of a routine clinical examination in most countries. Consequently, clinical gait analysis remains controversial.

Maturation of walking in children

A child's gait is different from an adult's until adolescence. The toddler walks with wide, short steps. The foot strikes the ground with the whole sole. Stance phase knee flexion is minimal. The legs are in external rotation throughout the swing phase. Reciprocal arm swing is absent. Stance phase is longer in young children compared with swing phase. There is increased muscle activity.

The gait pattern matures as the child grows older [A]. Heel strike begins at approximately 3 years of age. Stance phase knee flexion and external rotation values approach normal limits. Step width narrows and reciprocal arm movements begin at approximately 4 years of age. Cadence, step length and speed reach adult values at approximately age 15 years.

Longitudinal analyses of gait are necessary because of the tendency for gait patterns to change over time, and especially during growth.

Types of gait in diplegic and ambulatory total body involved children

Stability in stance, progression and foot clearance in swing are necessary for efficient walking. Stability is disturbed in CP because of impaired balance, increased muscle tone leading to contractures and muscle weakness. The common problems in stance are equinovarus, jump knee, crouch knee and internal rotation of the legs. Progression of the body is disturbed because of weakness and contractures as well. The common problems of swing are shortened step length and impaired foot clearance such as that which occurs in stiff knee gait.

The child's walking pattern changes with age. Diplegic children begin standing with the hips, knees and ankles extended and the legs crossed. Later, hip and knee flexion and ankle plantar flexion occur. Crouch occurs as the child grows older. Walking patterns are established at approximately 5 to 7 years of age.

In the sagittal plane, look for three types of pathologically abnormal gait: The jump, the crouch and the stiff knee gait.

Jump gait

The child walks with hips in flexion, knees in flexion and ankles in plantar flexion as if getting ready to jump [B]. This is typical for diplegic and ambulatory total body involved children when they begin to walk. The reason is spasticity of hip and knee flexors and ankle plantar flexors.

Crouch gait

Increased knee flexion and ankle hyperdorsiflexion occur during stance phase [C]. They occur in older children and after isolated triceps lengthenings that have been performed without addressing the spastic hamstrings. Hip flexors and hamstrings are tight, and quadriceps and triceps are weak.

Stiff knee gait

Decreased knee flexion occurs during swing phase [D]. The rectus femoris muscle is spastic and does not allow the knee to flex in initial and midswing phases. Limitation of knee flexion causes difficulty in foot clearance and stair climbing.

These sagittal plane gait patterns coexist with frontal and transverse plane pathologies. Look for scissoring and trunk

lurching in the frontal plane.
In the frontal and transverse planes look for scissoring gait and trunk lurching.

Scissoring gait and internal hip rotation
Scissoring gait is defined as crossing over of the legs during gait [E on previous page]. The cause is hip adductor and medial hamstring spasticity combined with excessive femoral anteversion.

Trunk lurching
Trunk lurching is an increase in the side-to-side movement of the trunk during walking [A]. It is caused by deficiency of balance. It may become worse after surgery and during periods of rapid growth.

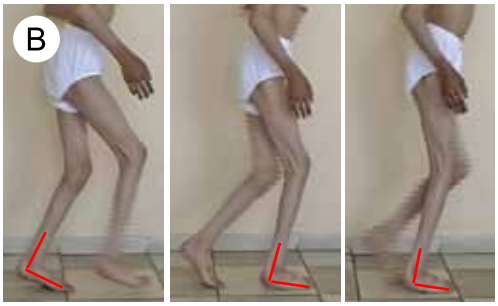
Traps to avoid: Apparent equinus
The cause of toe walking may not be gastrocnemius spasticity, but rather insufficient knee extension in certain children. When the patient is unable to extend the knee because of hamstring spasticity or knee flexion contracture, he or she seems to walk on tiptoe which can be mistaken for pes equinus.

Types of gait in hemiplegic children
Hemiplegic gait is subdivided into four types. With type 1, no active dorsiflexion of the ankle is present, and the foot in equinus. With type 2, a functioning tibialis anterior is present, and the foot is still in equinus because of the spasticity in gastrocnemius. With type 1, even if the gastrocnemius muscle is lengthened, the patient still needs a brace to keep the foot in neutral; however with type 2, lengthening of the gastrocnemius results in a more functional gait because the patient is able to dorsiflex the ankle. The differentiation between the two types of gait can be made using dynamic electromyography, which shows the activity in the tibialis anterior. With type 3, abnormal hamstring or rectus femoris activity is present, causing genu recurvatum or stiff knee, in addition to the problems observed with types 1 and 2. With type 4, in addition to the abnormal knee muscle activity, increased hip flexor and adductor spasticity or contracture are present.

Transverse plane deformities such as tibial torsion and femoral anteversion also might be present [C].

In spite of all technological advances in computerized gait analysis, certain gait abnormalities in CP continue to present difficulties for the clinician. The hints presented in the table help make better decisions for treatment [D].

The only remedy for trunk lurch is using a mobility device such as a walker or canes. Strengthening the hip abductors may also be helpful.



Distinguish apparent equinus from true equinus. Some children appear to walk in equinus but their ankle is actually in neutral or even dorsiflexed. Hamstring spasticity causes dynamic knee flexion deformity and the child walks as if he has equinus.

Femoral anteversion leads to intoeing and causes equinus. This type of deformity can also occur in hemiplegic children.

Hints on how to analyse gait D
Be familiar with normal child gait, watch children walk
Watch the child many times in different conditions
Record walking with a video camera
Ask the parents for photos and videos recorded at home and outside
Interpret gait data cautiously
Test balance and stability
Test in real life situations (at school, on the street)
Test speed

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A	Factors affecting prognosis
	Reflexes
	Absence of Landau, parachute sign
	Presence of Moro, ASTNR, STNR
	Timing of achievement of developmental landmarks
	Severity of involvement by the GMFCS
	Degree of intellectual involvement
	Sensory function, perception
	Motivation to move, interest to explore
	Family compliance
	Environmental factors
	Language/communication
	Body awareness
	Praxis (the planning and execution of movement)
	Behavior/cooperation
	Medical problems

B	Good prognosis for independent walking	Poor prognosis for independent walking
Head control	by 9 months	none by 20 months
Sitting	by 24 months	none by 48 months
Floor mobility	by 30 months	none by 48 months

Gross Motor Function Classification System (GMFCS)	
Level	*Ability* C
1	Walks without restrictions
2	Walks without assistive devices but limitations in community
3	Walks with assistive devices
4	Transported or use powered mobility
5	Severely limited dependent on wheelchair

Diplegic young child with good walking potential (GMFCS 3)

Mixed total body involved 13 year old child (GMFCS 5)

Prognosis and Goals of Management

The clinical picture in CP ranges from very mild to very severe depending on the extent of the CNS lesion. This wide spectrum of clinical findings makes it difficult to predict prognosis. Predicting prognosis forms the basis of management. The parents want to know about the prognosis of their child in order to shape their lives in the years to come. The physicians and therapists need to determine the prognosis in order to make sound treatment decisions.

The natural history

The brain lesion is nonprogressive and cannot be cured; however, the clinical picture changes as the child grows. This change is caused by the growth and maturation of the CNS and the musculoskeletal system. A small percentage of children who show signs of neuromotor developmental delay in infancy gradually outgrow their impairments. Others are left with neurological impairments ranging from mild to severe. Early intervention programs with exercises directed to minimize the neurological impairment exist, but because of the changing nature of the condition and the potential for the CNS to heal, it is not possible to tell whether the improvement is attributable to therapy or the ongoing CNS development.

Predicting functional prognosis

Clinical findings established over time help determine prognosis [A]. Potential ambulators usualy begin to walk between 2 to 7 years of age. Approximately 85% of partially involved children have the potential to become independent ambulators compared to only 15% of severely involved. Certain criteria help the physician determine prognosis in the young child [B].

Some major events in motor control have to occur in order for a child to walk. He must be able to hold his head before he can sit, and he must be able to sit independently before he can walk on his own. In children between 5 to 7 years of age it is easier to determine prognosis. The child with severe developmental delay who can not stand by age 5 to 7 is not going to walk.

Spastic hemiplegic and diplegic children with good cognitive function generally become independent walkers and productive members of the community. Most spastic hemiplegic children are able to become independent adults even without therapy. Physiotherapy, bracing, and efficient spasticity management result in a more efficient gait with less contracture formation in diplegics [D]. However, most of them still need orthopaedic surgery in childhood or adolescence. Approximately 85% of total body involved children [E] are unable to walk even indoors. They remain fully dependent on a caregiver and require assistive devices, special housing arrangements and lifelong continuous care. Physiotherapy, bracing, and drug treatment do not result in functional gains in athetoid or dystonic patients. Mild dyskinetic children use assistive devices and mobility aids to ambulate, and severely involved children remain totally dependent.

The Gross Motor Function Classification System is useful after age 2 years to determine prognosis [C]. The children in GMFCS I, II, and III are expected to walk with different levels of competencies. The goals for this group include the development of skills such as standing, walking, running and jumping, improvement of the efficiency of gait and maintenance and prevention of deterioration in gait. The children in GMFCS levels IV and V will have limited abilities to move around at

home, at school and in the community. Mobility and self-care goals are for sitting and comfortable positioning, transfers, dressing, changing diapers and – in general - maximizing the interaction with the environment. Improving general health status and decreasing medical complications have a high priority in this group.

Growing up with CP

'The child with CP grows up to be an adult with CP.'

As the disabled child grows into an adolescent and an adult, he will have to face problems integrating into the society he lives in. Disabled adolescents are deeply conscious of their body image and become psychologically vulnerable as they realize their differences from their able bodies peers. Rapid growth and increase in body weight at this age complicate the physical impairments.

Dealing with lifelong disability is mentally, spiritually, and financially exhausting for the family and the growing individual. Mental retardation, severity of disability, prolonged therapy programs preventing integration into mainstream education, overprotection of the adolescent and denial of disability by the family also have a negative impact on the prognosis for independent living in the adult [A].

Life expectancy

Mortality in CP is extremely variable. Life expectancy is normal in most diplegic and hemiplegic children who receive adequate medical care and have strong family support. Some severely affected quadriplegics die of malnutrition, infections or respiratory problems before reaching adolescence.

Treatment team

Many impairments of sensation, perception, cognition, communication, behavioural problems, epilepsy, difficulties with sleeping, drooling and feeding accompany the motor disorder seen in CP and require medical management. Therefore, health care for people with CP requires the skills of a variety of professions who must work efficiently and collaboratively with the family as a team. Each professional, and the teams involved in managing different issues (e.g. orthopaedic, epilepsy, feeding etc) must understand the range of problems associated with the condition.

The large team of experts consists of physicians, surgeons and allied health professionals such as physiotherapists, occupational therapists and child development professionals [B] who apply various treatment procedures [C]. The team must work together in harmony for success. The members must be aware of all the needs and limitations of the child and know what each other is doing for the child. The team should work also in cooperation with non-governmental organizations to support the integration of disabled children with society including sport, recreation and fun. Close interaction of the team members is vital [D]. Physicians and health professionals who work with patients with CP need to be aware of the implications of the diagnosis, the hardships these children undergo, and the value of each step gained towards function, no matter how small. The ultimate goal in the management is to minimize disability while promoting independence and full participation in society. Direct all efforts to gain independence in activities of daily living, ability to go to school, earn a living and a successful integration with the community.

A Factors adversely affecting independent living in the adult

Mental retardation
Severity of disability
Prolonged therapy preventing socialization
Overprotection of the adolescent
Denial of disability

B Treatment team

Physicians
Pediatric neurologist
Pediatric orthopaedic surgeon
Pediatric physiatrist
Allied health professionals
Physiotherapists
Occupational therapists
Orthotists
Psychologists
Speech therapists
Consulting members
Pediatric neurosurgeon
Ophthalmologist
Ear-nose-throat specialist
Audiologist
Dentist
Pediatric gastroenterologist
Nutrition specialist

C Treatment modalities

Support growth and nutrition
Ophthalmologic therapy
Rehabilitation
Glasses
Surgery
Dental hygiene
Gastrointestinal problems
Medications for reflux
Gastrostomy
Anti-reflux surgery
Therapy for motor function
Physical therapy
Occupational therapy
Adaptive seating
Bracing
Wheeled mobility
Orthopaedic surgery
Oromotor therapy
Chewing
Swallowing
Speech
Seizure prevention
Spasticity and dyskinesia
Medical treatment
Botulinum toxin
ITB, SDR

D

Teamwork is essential when treating children with CP. Even the best teams have controversies. Work close together and have educational activities which everyone participates in. Photo courtesy of D. Gaebler-Spira

Do not take too much of the child's time in therapy. Leave time for the child to play and socialize.

B

Treatment strategies		
Infancy	Supportive measures for prolonging and optimizing physical status and life	Nutritional support, exercise
Childhood	Maximum independent mobility	Medication, exercise, botulinum toxin, bracing
Preschooler	Maximum independent mobility, minimize deformity	Medication, exercise, botulinum toxin, bracing, surgery
Adolescence	Education, vocation and integration into the community	Schooling, sports, psychosocial support

As more children with CP come out in the open and participate in community life, people will begin to recognize them and make the necessary changes to allow the disabled to lead a normal life.

Teach activities of daily living.

The goals of health care for people with CP may sometimes conflict, for instance when strategies to limit deformity temporarily hinder activities and education. Share the rationale for any therapy intervention with the family and the individual with CP if possible to enable them to make an informed decision with the treatment team, to prioritise goals and to plan management regimens.

Treatment strategy

Everyone is a child only once. Do not sacrifice childhood for therapy [A]. Aim for a childhood that is as close to normal as possible. Base treatment strategy on a realistic evaluation of the child's present functional status and possible future prognosis. Set goals in each child and explain them to the family. Have the child participate in the goal-setting process when he is old enough. Individualize treatment for each child. Consider the child's age in the treatment plan [B].

Priorities in management are family education, addressing the specific needs of the infant, and providing adequate support for optimal growth and development. Play activities, attending mainstream education if possible, and socialization with peers play a crucial role in the development of the child. Plan therapeutic procedures such as physiotherapy and surgery carefully so as not to interfere with school and social life. Ideally, the society must be reorganized [C] to support healthcare providers, caregivers and families to enable integration of the individual with CP with the community.

Goals differ for children who can move around by themselves and bear weight on their lower extremities. The child who can move himself from one chair to another in the house, even if by crawling on the floor, is very different from the child who cannot move at all. The ability to move independently and bear weight on lower extremities is an indicator that the child will respond to aggressive treatment even if he has not received therapy before.

Explain all treatment procedures to the family and if possible to the child. Make sure they understand. Clinical findings, functional limitations and priorities change as the child grows. Continuously review and revise treatment plans for each individual child. This is the real challenge and joy of treating a child with CP.

Priorities in function / treatment (From E. Bleck)

Communication

Communication is necessary to express thoughts, feelings and needs. Every individual with CP needs a a way to communicate to be part of the family and the community. If the child can produce comprehensible sounds and syllables by 2 years of age, he will probably have normal verbal communication. Consider alternative communication methods such as simple communication boards or high technology equipment like computers in children who have difficulty speaking.

Activities of daily living

Activities of daily living are self-care activities such as feeding, toileting, bathing, dressing, and grooming in addition to meal preparation and household maintenance. Dyskinetic and total body involved children have problems of dexterity and fine motor control that prevent independence in activities of daily living. Hemiplegic and diplegic children can become functional in these areas [D]. They sometimes need help of occupational therapy. Family attitude is a critical factor determining the level of independence of a child. Overprotection results in a shy and passive individual who has not gained self-care abilities.

Mobility

Children have to explore their surroundings to improve their cognitive abilities. Mobilization is crucial for the young child with disability to prevent secondary mental deprivation. Use wheelchairs or other mechanical assistive devices to promote independent mobility in the community if the child cannot achieve mobility by walking.

Mobility is important to function in the fast paced societies we live in. People who have difficulty moving are always at a disadvantage. In the adult, becoming an independent member of the society and earning a living depend upon independent mobility [A].

Wheeled powered mobility allows the total body involved individual to participate in social life.

Ambulation

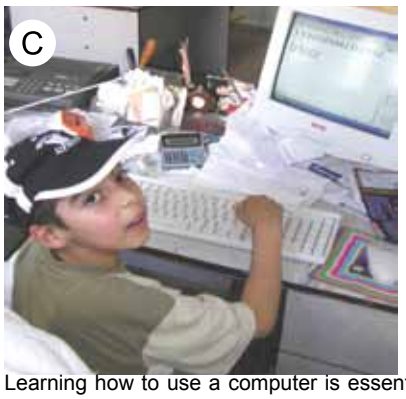
Families view ambulation as the most important issue during childhood. Every effort must be made to increase the child's ability to walk; however, walking depends more on the extent of the child's neurological impairment rather than the amount of physical therapy, surgery or bracing he receives. The child achieves his or her maximum potential with practice.

Priorities change in adolescence. The adolescent needs education [B], independence, and an active social life. For him, although still important, ambulation is the least needed function. Learning how to use the computer may benefit the adolescent more in the long run than being able to take a few assisted steps [C]. Mobility is important for the child, whereas social identity and independence are more valuable for the adolescent.

Quadriplegic children generally cannot enter mainstream education and have to receive special education.

Psychosocial issues

Children who receive intensive physiotherapy and surgery without apparent gains have psychological problems in adolescence and adulthood. Lack of independent mobility and presence of deformities in spite of prolonged years of therapy frustrates the adolescent who becomes aware of the difference between him and his peers as he grows older [D]. The child with CP grows up to become the adult with CP. He has to continue life as a disabled person facing physical as well as spiritual barriers of the society. Sometimes he is forced into tasks which his functional capacity will not allow and at other times he is barred from social life. Both result in increased frustration, anxiety neurosis or depression which decrease independence further. Keep all these problems in mind while formulating a treatment plan to address the individual's needs [E].



Learning how to use a computer is essential in this century. Teach the child computer skills for better quality of life.

Talk to the adolescent alone. She may have things to tell you that she does not want anyone to hear. Try to understand her point of view.

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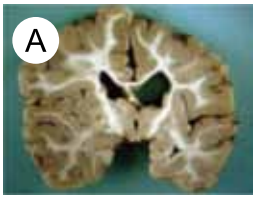
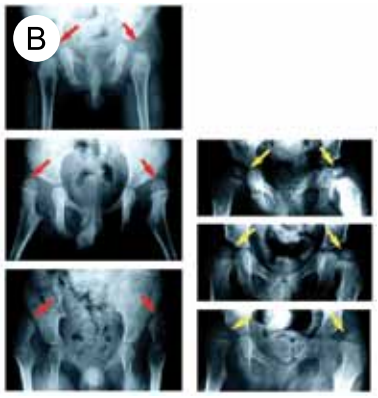
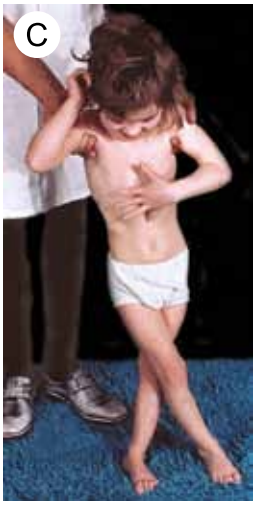
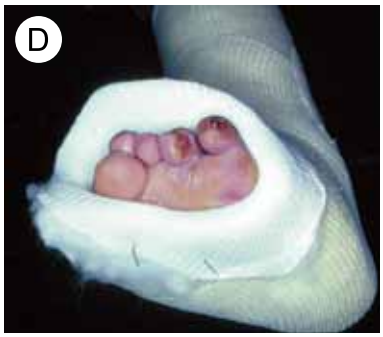
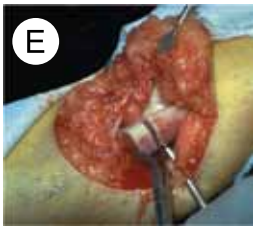
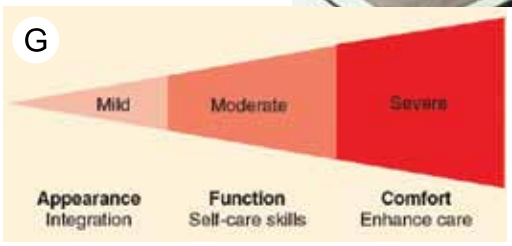












* This work was presented by Dr. Lynn Staheli at the 2nd Eastern Mediterranean Cerebral Palsy and Developmental Disorders meeting in 2004 in Santorini, Greece.

Management Principles In Neuromuscular Disease Based On Valuing Childhood

Courtesy of Lynn Staheli MD and Lana Staheli PhD *

When surveyed, most people rate childhood as the most happy and valuable part of their life. Childhood has intrinsic value, it is not a time to be sacrificed in preparation for adult life. The childhood of children with CP is often placed at risk because it is squeezed out by treatments. Health care professionals have an enormous influence on the life of the child and the family. Management principles help keep a balance between medical interventions and preserving childhood. Over the past four decades we have found these twelve principles useful in maintaining this balance.

1. Consider the natural history of the disorder.

The problem is a neurological lesion [A]. The effects of this lesion include motor, sensory and integration difficulties. Tertiary problems include dynamic deformities of the musculoskeletal system that tend to become fixed with time. Fixed contractures cause altered loading of joint cartilage, disturbed growth and bony deformity [B]. These deformities limit function and mobility, and eventually cause degenerative arthritis and pain. Understanding this sequence is important in planning management and preventing adverse outcome. A knowledge of natural history helps us differentiate the effect of our treatment from that of growth and maturation of the child.

2. Appreciate the significance of sensation and perceptive disabilities.

The child with cerebral palsy has a loss of sensation that is not often appreciated. A diagnosis of spastic diplegia [C] does not acknowledge the existence of any sensory component. In the child with hemiplegia hand function may be more limited by the sensory loss than the deformity and muscle weakness. The child with arthrogryposis with severe deformity still functions well because of intact sensation. Skin ulcers are common in children with myelodysplasia [D].

3. Recognize the limitations of treatments.

Our treatments do not correct the primary neurological lesion. Our inability to cure the disease means we manage symptoms or deformity [E] Acknowledging these limitations is important in developing a treatment plan that balances time for treatment and time for being a child.

4. Be cautious with comparisons.

Our objective is to give the child the best possible life-not to make the child normal. Be cautious about using normal values to assess children with cerebral palsy. Becoming too focused on making limbs straight or gait lab curves normal [F] may be counterproductive.

5. Focus on appearance, function and comfort, not on deformity.

Focus management to the individual's needs. Base management principles on severity of the problems [G].

6. Provide functional mobility.

Provide functional mobility to promote intellectual and social development. Functional mobility must be practical, effective and energy efficient. Walking is only one method of mobility. If necessary, provide mobility aids early to increase independence [H]. Children do not become addicted to these aids. Make time for exploration.

7. Establish appropriate priorities.

Adults with CP rank communication and socialization [A on next page] above mobility in importance. Frequently the family's major

concern is whether or not their child will walk. Walking is important but not essential.

Our goal is to help family understand that the most important objectives are independence, social integration and mobility-not necessarily walking. During each visit attempt to keep the focus on these long term objectives. Help the family accept the limitations caused by the neurological lesion and the reality that this cannot be corrected. Avoid becoming focused on minor problems that consume energy and resources without long-term value. Make time for friendships with other children.

8. Focus on the child's assets.

Like other children, children with disability have talents that need to be identified and developed. Take every opportunity to compliment the child and the parents. Time spent developing the child's assets [B & C] is usually more productive than time spent attempting to overcome the child's disability. Make time for creativity.

9. Shift priorities with age.

In early childhood focus on mobility [D] and self care. In mid childhood focus on socialization and education. In late childhood focus on vocational preparation. Make time for just fooling around.

10. Maintain family health.

Protect the health and well being of the marriage and the family. Help the family and support groups to provide information, perspective, support and friendship. Recognize that all treatments have a cost to the child and the family. Avoid overwhelming the family [E]. Make time for family fun [F]. The family is like a computer. If too many programs are opened at once it will crash. Monitor the family's stress and avoid overloads.

11. Avoid management fads.

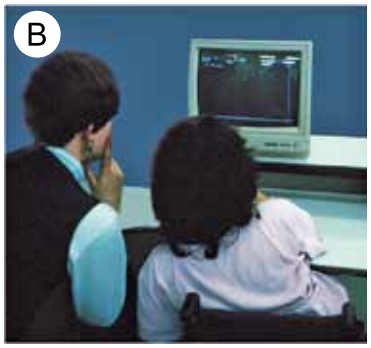
History of medical management includes a vast number of treatments that were either harmful or ineffective [G]. Children are vulnerable, adults would never tolerate what has been done to children. Steer the family away from interventions that are unproven or unrealistic. Such treatments drain the resources of the family and lead to eventual disappointment for the child. Extensive bracing, misguided operations and exhaustive therapies are examples of treatments once in vogue but later abandoned. Often management methods are like waves, a rise and fall followed a new wave of some new treatment. We cannot cure these disorders but we can care for the child and the family. Most important- care not cure.

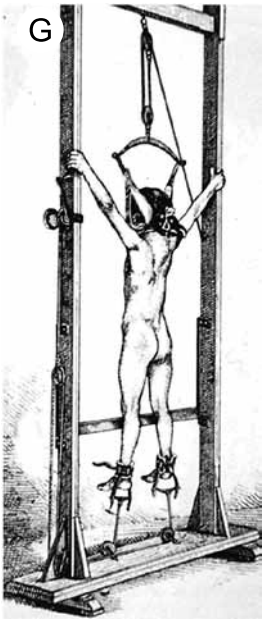
12. Protect the child's play experience.

The objective is a child who meets his potential both emotionally and physically. Play is the occupation of the child [H]. The child with a disability needs to play just as other children, perhaps more. Preserve time and energy for this experience. The individual is a child only once. Special Olympics and wheelchair basketball are examples of appropriate team sports. Spontaneous play is best. Let the child discover the joy of childhood. Monitor the child's time and preserve time for play.

Summary

The relationship with the health care provider is very important in the child's life. Monitor and preserve the health of the family. Avoid excessive stresses by too many programs. Help the family accept the child's problem. Compliment and affirm the child and the family whenever possible. Focus on the child's assets. Provide a time for childhood with play.





A

Goals of rehabilitation	
Improve mobility	Teach the child to use his remaining potential
	Teach the child functional movement
	Gain muscle strength
Prevent deformity	Decrease spasticity
	Improve joint alignment
Educate the parents	To set reasonable expectations
	Do the exercises at home
Teach daily living skills	Have the child participate in daily living activities
Social integration	Provide community and social support

B

Components of rehabilitation
Physiotherapy
Occupational therapy
Bracing
Assistive devices
Adaptive technology
Sports and recreation
Environment modification

C

Mobility is essential for successful integration into the community.

D

Rehabilitation planning
Example: Independent standing
1- State the necessary time period to achieve this goal i.e. 12 weeks
2- Plan the methods to achieve this goal i.e. Stander, exercises to improve trunk balance
3- Evaluate the end state.
4- Revise the treatment program or the goals if not successful.

Rehabilitation & Physiotherapy

Rehabilitation is the name given to all diagnostic and therapeutic procedures which aim to develop maximum physical, social and vocational function in a diseased or injured person. The goal of rehabilitation is to gain independence in activities of daily living, school or work and social life. This is possible to the extent of the person's impairments.

Components of child rehabilitation

Child rehabilitation consists of improving mobility, preventing deformity and educating the parents about the child's problem [A]. It also involves helping the child to learn the skills he will need in daily life, school and while playing with friends. Lastly, rehabilitation means decreasing the complications which arise as a result of the child's neuromuscular impairments.

Therapeutic exercises help the child learn how to sit, stand, walk and use his upper extremity for function. The child also learns how to use his remaining potential to compensate for the movements he cannot perform. Decreasing spasticity, gaining muscle strength and improving joint alignment decrease deformity. The education of caregivers involves gently coaching them to set reasonable expectations for their child, and teaching them to follow their child's exercises at home. Parents should encourage their children to participate in daily living activities by using the functional skills they learned during therapy. Community and social support is another aspect of rehabilitation.

There is no method which can decrease the neurological impairment. Explain to the parents not to spend valuable time and hope with alternative treatments. Aim to have the child fulfil his maximum physical, intellectual and psychological capacity and have a happy childhood as close to normal as possible. Focus on the child's abilities and interests. Try to improve function by working on these. The child can easily improve in the activities he likes doing. This will enable him to have a happy childhood and a job in the future.

CP rehabilitation [B] consists of physiotherapy, occupational therapy, bracing, assistive devices, adaptive technology, sports and recreation.

The main aim of rehabilitation is providing an education for the child, and to help him grow up to be a productive, independent adult. Various therapy procedures exist some of which do not really relate to real world situations. The skills that the child gains during therapy sessions should be useful within the community. Never ignore the child's education throughout the various therapy procedures. Always aim to send the child to school for an education and prepare him for community integration [C].

Planning rehabilitation

The child begins to receive physiotherapy when he is a baby. Occupational therapy starts towards age two to teach daily life activities. The toddler uses assistive devices for mobility. Bracing may be necessary as the child begins to walk. Sports and recreation are crucial for the school aged child. Play is important beginning in infancy throughout adolescence.

Have short and long term goals depending on the child's expected functional outcome [D]. Evaluate the child, specify these short and long term functional goals and set a time limit in which you expect the child to achieve these goals. Review the plan if the child cannot achieve the expected function in the predetermined time period.

Factors influencing rehabilitation outcome

Consider the following factors in the rehabilitation plan [A].

Treatment team

The pediatrician provides diagnosis and preventive health care. The orthopaedic surgeon tries to minimise static and dynamic contractures to improve mobility. The pediatric physiatrist evaluates the child's overall medical, surgical and therapy options and helps the child and the family to set functional, achievable goals. Together, the rehabilitation team works to assist the person with CP to achieve his place in the society. A productive interaction between the physicians and the therapists is essential for the maximum benefit of the child. All those involved with the child must have a basic understanding on the diagnosis, family expectations, degree of motor dysfunction, functional goals and the therapy program.

Medical problems of the child

The rehabilitation physician and the team must be prepared to anticipate certain acute and chronic problems during the rehabilitation of the child with CP. The disabled child is more prone than his able-bodied peers to respiratory problems, convulsions, dysphagia, depression, gastroesophageal reflux and sleep disorders. Total body and some severely involved diplegics have visual and hearing deficits, mental retardation, cortical sensory deficits and communication deficits that prevent the child from reaching his maximum potential.

The child's character

The motivation to move, temperament, behavior/cooperation and the willingness to take risks are important determinants of rehabilitation outcome. These personality characteristics of the child are independent of impairment or disability.

The family

Some families provide their children with ample experiences and opportunities that enrich their environment and increase their ability to achieve new skills. Family resources, quality of home environment, family support and parent/caregiver expectations guide the plans of long-term care for the disabled child.

Physiotherapy

Physiotherapy helps improve mobility. It is the basic treatment in all children with CP [B]. It consists of exercises, bracing and activities towards reaching specific functional goals. It aims to bring the child to an erect position, give the child independent mobility and prevent deformity. Organize physiotherapy to fit into the family's lifestyle.

There are numerous techniques available, with limited scientific basis to support their use. Optimize caution when recommending therapy modalities [C].

The 'dose' of physiotherapy intervention (e.g., frequency, duration, etc.) is often decided empirically, following tradition and modified by economic considerations; the dose is seldom evidence-based and therefore may not be optimal.

General principles of physiotherapy

Physiotherapy begins in early infancy and continues throughout adolescence [A on next page]. The primary purpose is to facilitate normal neuromotor development. With the help of correct positioning, appropriate stimulation and intensive exercise the therapist tries to gain head control, postural stability and good mobility in the child. This is possible only to the extent of the child's neurological capacity. Even with vigorous physiotherapy many children remain functionally impaired in varying degrees.

There are different methods of therapy for children with neurological impairments. Even though they differ in the techniques they use,

Factors influencing rehabilitation outcome — A

Treatment team	Productive interaction Basic understanding
Medical problems	Respiratory Convulsions Dysphagia Depression Gastroesophageal reflux Sleep disorders Visual and hearing deficits Mental retardation Sensation Communication
Child's character	Motivation to move Temperament Behaviour/cooperation Willingness to take risks
The family	Resources Quality of home environment Support Expectations

B

Physiotherapy tries to improve

- Postural control
- Muscle strength
- Range of motion
  - Decreasing spasticity and contracture
  - Increasing muscle elasticity and joint laxity
- Joint alignment
- Motor control
- Muscular/cardiovascular endurance and mobility skills
  - Increasing coordination / agility
  - Balance
  - Transitions
  - Use of assistive devices

According to the available scientific evidence, do NOT use:
An exercise program comprised solely of passive range of motion exercises delivered by a therapist. Parents, other caregivers or patients can be instructed to carry out these exercises themselves.
Passive reflexive techniques.
Sub-threshold Transcutaneous Electrical Nerve Stimulation (TENS) when the goal is to increase muscle function.
Classical Bobath/neuro-developmental therapy (NDT) where the emphasis is on "normalization" of muscle tone through passive handling techniques such as the use of reflex inhibiting patterns.
According to the available scientific evidence, recommend:
Exercise activities that include active participation of the patient which include functional goals.
Progressive Resistance Strength Training.
Constraint Induced Movement Therapy in the upper limb.
Use of assistive technology to promote mobility such as orthoses, wheelchairs, walkers or crutches.
The efficacy and effectiveness of the following are uncertain, evaluate carefully:
Partial Body Weight Supported Treadmill training
Robotic assisted walking (e.g. Lokomat®)
Night splinting

C

Therapy program	
Infant	Stimulating advanced postural, equilibrium and balance reactions to provide head and trunk control
Toddler & preschooler	Stretching the spastic muscles, strengthening the weak ones, and promoting mobility
Adolescent	Improving cardiovascular status

A

Principles of therapy methods
Support the development of multiple systems such as cognitive, visual, sensory and musculoskeletal
Involve play activities to ensure compliance
Enhance social integration
Involve the family
Have fun

B

Basic problems in the neuromotor development of children with CP
Difficulty with flexing and extending the body against gravity
Sitting
Functional ambulation

C

For functional ambulation a child needs
Motivation to move
Enough muscle strength and control
Able to shift his body weight (balance)
Awareness of body position and movement (deep sensation)
Sufficient visual and vestibular system
No deformities interfering with joint function

D

basic principles remain the same [B]. The problems of neuromotor development are difficulty flexing and extending the body against gravity, difficulty sitting and functional ambulation [C].

For functional ambulation a child needs motivation to move and explore the world around him [D]. He must have enough muscle strength and control. He must be able to shift his body weight and have an awareness of body position in space at rest and during movement. Visual and vestibular systems must be sufficient. There must not be any deformities interfering with joint function.

In physiotherapy sessions the therapist works with the child in supine [E,F] and prone [G,H] positions to improve head and trunk control. She supports the child in the sitting position [I,J] to develop weight shifting and unilateral balance, ability to rotate the body and the ability to respond to sudden changes in position. The rehabilitation team strives for long-term, functional mobility in a variety of environments so that the child will integrate into the community and social life in a healthy way.

Therapy methods

Stretching, range of motion and strengthening exercises are essential in all children. In addition, neurofacilitation techniques stimulate the central nervous system to establish normal patterns of movement. These neurofacilitation techniques were developed over the years to minimise the neurological impairment and help the healing CNS to reorganize. This has not been possible and the focus of therapy shifted from trying to heal the neurological lesion to increasing motor function. There is no treatment method that can heal the lesion in the CNS. The intact neurons in the brain may substitute for lost function, new synapses may form and reorganisation of neurons take place so that the child gains function as he grows. This process is termed neuronal plasticity. The present neurofacilitation methods stimulate the CNS and accelerate neuromotor maturation through the process of neuronal plasticity. The Vojta method is common in Eastern Europe whereas the neurodevelopmental training technique established by B. Bobath and named after her is widely used in the Western world. Both because of difficulties in diagnosing CP in infancy, and the inherent potential of the CNS to heal, it is extremely difficult to judge the actual success of such procedures.

Flexion against gravity promotes visual and orientation, self-touch/ body awareness and active hip mobilization. It is preparation for rolling/floor mobility.

Support in prone facilitates visual orientation, head control, development of postural extension and active control of weight shift.
Photos courtesy of S. Sterchi

Children who use the upper extremities for support when sitting can not develop hand function. Provide a seating support for better head and trunk control and give the child the opportunity to play. This way, he will have better vision, use his hands actively and have improved breathing capacity due to proper vertebral alignment and chest support. The child can also shift his body weight over the pelvis when supported in this position to exercise trunk rotation.

Plan the exercises according to the mobility needs of the child. It is not enough to have a therapy session only once during the day with the therapist. Children with CP need to exercise at home to gain maximum function. The success of the techniques used in physical therapy depends on repeated practice. The parents must repeat the exercises with their children every day and observe children for improvement or changes that may be needed.

Conventional exercises

Conventional exercises consist of active and passive range of motion exercises, stretching, strengthening and fitness to improve the cardiovascular condition [A].

Range of motion [B] and strengthening exercises [C] continue for a lifetime in CP, but are especially important in the recovery stage following orthopaedic surgery and in the growth spurt period when risk of contracture formation is maximal. Teach the stretching exercises to the caregiver to prevent contractures. Physiotherapists and parents should be careful with stretching exercises so as not to tear muscle fibers and cause pain, otherwise they will have the opposite effect. Night splints and stretching are not by themselves sufficient to prevent contractures. Strengthening exercises to the antagonist muscles are always necessary because spastic muscles are also weak. Sports activities are helpful in decreasing stiffness and contractures in adolescents using the wheelchair. Cardiovascular conditioning is crucial for the total body involved individual in the wheelchair.

Balance is a prerequisite for independent walking, balance training is one of the key reasons for physiotherapy [D,E].

Strengthening [F] does not affect muscle tone, it does not increase spasticity. On the contrary, the importance of strengthening the spastic muscles and their antagonists cannot be over emphasized for efficient motor function.

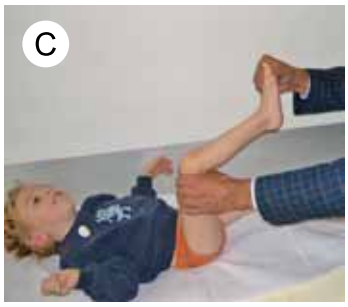
Do not prevent sitting in the W-position [G] for fear of hip subluxation. W-sitting does not increase femoral anteversion or cause hip subluxation. Children with femoral anteversion sit in a W-shaped position because it is comfortable. When forced to change position for fear of contracture, the child needs to use his hands for balance which interferes with hand function.

Neurofacilitation techniques

Sensory input to the CNS produces reflex motor output. The various neurofacilitation techniques are based on this basic principle. All of the techniques aim to normalize muscle tone, to establish advanced postural reactions and to facilitate normal movement patterns.

Vojta method of therapy Vojta established 18 points in the body for stimulation and used the positions of reflex crawling and reflex rolling. He proposed that placing the child in these positions and stimulation of the key points in the body would enhance CNS development. In this way the child is presumed to learn normal movement patterns in place of abnormal motion. Positioning and stimulation techniques are different from NDT [A, B on next page]. Vojta states that therapy should be applied by the primary caregiver at home at least 4-5 times daily and stopped after a year if there is no improvement.

A	Conventional exercises
	Active and passive range of motion
	Stretching
	Strengthening
	Fitness



Conventional exercises are an integral part of physiotherapy. Stretching exercises are helpful for contractures whereas strengthening is necessary for weak muscles.

Equilibrium reactions and balance can take a very long time to develop and sometimes do not develop at all despite intensive training.
D: Courtesy of M. Sussman

Strengthening is essential for function. It does not increase spasticity or cause deformity.

Do not prevent W sitting if the child is comfortable and relaxed in this position. It is only a result of increased femoral anteversion and will not cause hip subluxation.


In the Vojta technique the therapist positions the baby to initiate crawling and rolling. The principle is to hold the head in a straight line with the body. Stimulating certain points in the body by applying pressure with the fingers causes reflex movements. It is unclear whether these reflex responses evolve into voluntary movement in the child.


Therapists try to elicit advanced postural reactions while preventing abnormal movements in the Bobath approach. D : Courtesy of G. Koloyan

E What can physiotherapy accomplish?
Assist in the neurological development of the child.
Enable the child to use his existing potential in the best possible way.
Improve mobility and prevent secondary psychosocial retardation.
Prevent contractures and deformities in some cases.


Therapists try to develop balance in the kneeling position. Development of the control of pelvic muscles is an important step.


Work on ambulation in the parallel bars and with crutches. Use a wedge on the floor to keep the feet apart.

Bobath neurodevelopmental therapy This is the most commonly used therapy method in CP worldwide. It aims to normalize muscle tone, inhibit abnormal primitive reflexes and stimulate normal movement. It uses the idea of reflex inhibitory positions to decrease spasticity and stimulation of key points of control to promote the development of advanced postural reactions [C]. It is believed that through positioning and stimulation, a sense of normal movement will develop. An important part of therapy of the infant is teaching the mother how to position the child at home during feeding and other activities [D]. The baby is held in the antispastic position to prevent contracture formation.

Benefits and limitations

Physiotherapy cannot correct the movement problem in CP. A few rare cases reach their full potential through physiotherapy alone, the majority of children need other interventions. The effect of physiotherapy in preventing contractures and deformities or improving balance and coordination [F,G] is also limited. Physiotherapy is beneficial in promoting the neurological development of the child and teaching the child to use his existing potential in the best possible way. By improving mobility, physiotherapy may also prevent secondary mental and psychosocial retardation. However, the success of treatment depends on the neurological capacity of the child. An allegory can be made with sports: even with the best coaching, an athlete cannot compete in the Olympics if he does not have the potential. Similarly, even with the best physiotherapy, the child with CP cannot walk [H,I] if he does not have the neurological capacity. The treatment team must be careful therefore not to raise any false hopes about the outcome of physiotherapy in children.

The efficacy of neurofacilitation techniques in improving the neurological impairment is controversial. Meta-analyses of neurodevelopmental therapy (Bobath) have shown that the functional status of the children at school age are the same regardless of having received therapy or not.

Two most important aspects of physiotherapy in children are strengthening and functional activity. Direct efforts to strengthen the weak spastic muscles and to teach children functional activites.

Long hours of intensive physiotherapy can harm the child in many ways. It interferes with play, schooling, family and peer relations. Organize therapy so as not to disturb normal childhood.

Occupational therapy and play

OT aims to improve hand and upper extremity function in the child through play and purposeful activity. There are defined systematic treatment methods for occupational therapy. Sensory integration therapy aims to enhance the child's ability to organize and integrate sensory information. In response to sensory feedback, CNS perception and execution functions may improve and the motor planning capacity of the child may increase. Constraint induced movement therapy where the normal hand is constrained and the paralytic hand is forced to function may be useful in children with hemiplegia.

Begin therapy toward one year of age when the child can feed himself using a spoon and play with toys. Teach the child age-appropriate self care activities such as dressing, bathing and brushing teeth. Encourage the child to help with part of these activities even if he is unable to perform them independently.

Always include play activities in the rehabilitation program [A, B]. Play improves mental capacity and provides psychological satisfaction. Organized play can address specific gross and fine motor problems in the child and take the place of boring exercise protocols. This increases the child's compliance with therapy. For example, riding a toy horse may improve weight shift over the pelvis, swinging may improve sensation of movement.

Sports and recreation

Disabled children need to be involved in sports and recreational activities just like their able bodied peers. Sports and recreational activities also form part of the rehabilitation program. Physical activity plays an important role in physical development, general fitness and health. It provides fun and recreation. Physical and occupational therapy combined with recreational activities or adapted physical education increases efficiency of rehabilitation and assist the disabled child to use his potential. It is difficult and time consuming for a child to continue physiotherapy once he is in school. If he does, he will be alienated from his friends. Sports and recreation benefit the child because they save him from going to long hours of physiotherapy and being apart from his friends, from loss of valuable time for lessons and play. In the meantime sports provides the only means of improving the child's neuromotor abilities and preventing deformities when he is at school. Through sports and recreational activities, the child has the ability to participate in the world of normal children, will not feel left out because of hours of physiotherapy and will improve his neuromuscular functional status.

There are a variety of summer and wintertime sports that the disabled child can participate in [C,D]. The competitive aspects of involvement in sports and games is basic to human nature, however many physically handicapped individuals frequently do not have the physical attributes to participate fully in different sport activities. Therefore most of the modern games and sports are modified for handicapped persons. The rules of the sport or the game are modified to meet the needs of disabled person (wheelchair tennis), or specially designed adapted devices are used for physically disabled to compensate or substitute the loss of muscle strength or function (downhill skiing) [E]. Some modern sports require very high-technology equipment, but most adapted equipment and devices can be easily made or adjusted locally [F].

Sports scientifically shown to have significant therapeutic effects in CP are swimming [G,H] and horseback riding. They help to increase muscle strength and range of motion in the joints, improve sitting balance and body control and provide fun. Swimming [A on next page] starts with basic pool exercises. In the whole body involved child, it helps normalize muscle tone and decreases rate of contracture. In hemiplegic and diplegic children, it can strengthen muscles, improve cardiovascular fitness and efficiency of gait.

Horseback riding [B on next page] improves head control and trunk balance, normalizing muscle tone. It creates a lot of positive emotions in interaction with the horse. It also improves self esteem in the child [C on next page].

The child develops hand-eye coordination by playing with simple toys. She also experiences a sense of accomplishment, improving self image.

Children can learn simple tasks at home. They can learn how to look after themselves. They need coaching to master bathing, grooming and toileting skills.

Wintertime sports
Downhill
Two track skiing
Three track skiing
Four track skiing
Mono-and bi-skiing
Sit-skiing
Cross-country skiing

C

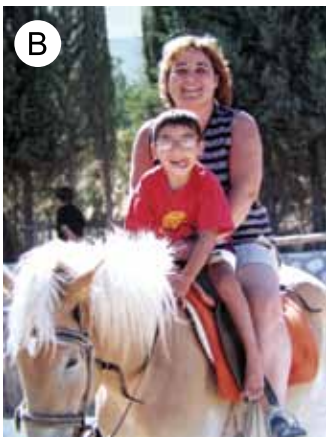
Summertime sports
Swimming
Athletics
Wheelchair basketball
Tennis
Table tennis
Wheelchair racing
Shooting
Archery
Canoeing
Kayaking
Sailing
Weightlifting
Football

D

E & F courtesy of Shriner's Hospital, Portland

Most children love to swim, or at least be in the water. The buoyancy of water helps movement and inhibits muscle tone. Splashing about is a lot of fun. Prescribe aquatic exercises as therapy in the less severely involved child.

A	Advantages of swimming
	Normalizes muscle tone
	Decreases rate of contracture
	Strengthens muscles
	Improves cardiovascular fitness
	Improves walking



Riding a horse is beneficial for the child in many ways. It gives the child self confidence, a feeling of responsibility, improves balance and posture.

C	Horseback riding
	Improves head control
	Trunk balance
	Normalize muscle tone
	Positive emotions
	Self esteem

	Recreational programs
D	Arts and crafts
	Music
	Dancing
	Wheelchair dancing
	Drama, camping
	Fishing
	Scuba diving

'Bridge of Hope - Mobile Theatre' was founded in Armenia. It is a drama group in which disabled children act and have a chance to express themselves in art.

Courtesy of G. Koloyan

Teach the parents to take care of the baby. Infancy is a time for parent-infant bonding.

Even vigorous sports activities do not cause or increase orthopaedic deformity. In developed countries adapted physical education and sport are included in public school programs. In developing countries adapted sport and recreation [D] is organized by special NGO's (non-government organizations) or sport organizations. The Bridge of Hope from Armenia is a good example [E].

Rehabilitation strategy and goals according to age

Rehabilitation aims to prevent disability by minimising the effects of impairments, preventing secondary disabilities and maximising motor function throughout the child's life. The focus of treatment shifts over the years but the principles remain the same.

Functional goals change as the baby becomes a child and the child matures into an adult. Younger children focus mainly on mobility whereas adults shift their focus to communication and activities of daily living. The first four years are spent in physiotherapy and bracing, orthopaedic surgical procedures are performed between 5 to 7 years of age, education and psychosocial integration become main issues in the school age (7-18 years).

Reach mobilization goals by the time the child is ready to go to school. The child needs aggressive physiotherapy in the growth spurt period and after orthopaedic surgery. Communicate with the school physiotherapist or the physical education teacher to enable an active life for the child at school.

Early intervention

Early intervention is the general name given to many therapy modalities including exercise and caregiver education. Early intervention programs involving infant stimulation and caregiver education [F] may retard or reverse the central nervous system lesion causing the clinical picture of CP and thus prevent or minimise neuromotor delay. There is no established routine and no proven value of these programs, however until we know which babies are going to be normal on their own, it is better to let them have the benefit of early treatment so that any improvement potential is not lost. Despite the controversies early treatment benefits the parents. They receive a great deal of practical advice and support this way. The child's functional status may improve with parental support. Early treatment creates more opportunity for the potential to develop any normal abilities and for decreasing the defects.

Infancy

Rehabilitation goals are to educate the family about the child's problem, to improve parental bonding, to help the mother care for the baby and to promote optimal sensorimotor development through positioning, stimulation and exercises [A on next page] if possible. Increase mobility and help the baby explore his surroundings.

Use positioning, carrying, feeding and dressing techniques which promote bodily symmetry in the infant. These limit abnormal posture and movements and make functional activity possible. Provide sensory stimulation using various movements and postures. Some positions lengthen the spastic muscles and make voluntary movement easier. Add weight shifting, weight bearing, trunk rotation and isolated movements into the exercise regimen.

Customized seating or seating supports are necessary. Visual attention, upper extremity use and social interaction improve in the child who is supported in sitting. When the child is sitting comfortably without fear of falling he sees his hands, practices midline play, reaches for his or her feet and sucks on his fingers. All of these movements provide sensory stimulation and promote a voluntary motor response to that stimulation. Active movements encourage the infant to develop flexor control and symmetry. Use toys that require two hands, facilitate the use of neck and trunk muscles and anteroposterior control of the head. Educate the parents to help them accept their child's problems and raise their child in the best possible way.

A crucial part of therapy is teaching the exercise program to the mother. Parents must be accustomed to taking the active part in their child's treatment early on.

Childhood

The needs of the children are different starting at age one depending on the type and severity of involvement. Redefine goals of treatment at this stage. Limitations in motor function create disabilities in learning and socialization. The child cannot become independent. The major goal of rehabilitation in the preschool period is achieving independent mobility [B]. At this age child's maximum level of motor function can be predicted with greater accuracy. Choose a method of mobilisation and teach the child how to use it so that he will be free to explore his environment.

Bring every child to an erect position regardless of prognosis for walking [C]. Encourage the use of standers. Focus on independent mobility in every child. The total body involved child needs sitting supports and wheeled mobility. The diplegic and the hemiplegic must be supplied with appropriate bracing to begin to work on ambulation.

A very important component of rehabilitation is having fun.

As the child matures physically by the age of four he must be involved in self care activities and activities of daily living at home. Always aim for functional tasks in the limits of the child's capacity [D]. Address educational issues with help from special educators if necessary. Plan and complete all surgical procedures directed towards better ambulation by school age if possible.

School age and adolescence

Children in mainstream schools regress because of a lack of exercise. Physiotherapy done in clinics in contrast, takes too much time, causes separation of the child from his peers and prevents socialization. Physiotherapy should be performed at school if possible with the help of a community physiotherapist or the physical education teacher. Coordinate school with play and exercise. Handle social and vocational issues during school years for a better state of independence. Efforts to improve the psychological well being of the child are necessary especially in adolescence. Children with CP have a significantly lower level of physical activity and cardiovascular fitness compared to their healthy peers even if they are only mildly impaired. Keep in mind the fact that cardiovascular issues continue to be a problem for the CP patient in adulthood, increase the activity levels of these children at school by sports and play [A on next page].

The ability to stand and take a few steps should be preserved in the adolescent even if only for therapeutic purposes.

Therapy sessions must focus on meaningful activities such as climbing stairs.

One goal of therapy is to enable the child to play with peers is and participate in age-appropriate activities.

Functional gains may be few in many children with CP. Minimizing medical problems and comorbidities sustains family health and happiness. This ten year old girl with quadriplegic CP is happy among her large family who are well adjusted to her disability.

The family

Physical impairments that create lifelong disability for the child cause psychological disturbance in the family. The parents are in need of constant support, have problems understanding and accepting the situation and tend to blame themselves or the physicians. Explain the nature of the problem to the family and include them in the treatment plan. Remember that families may need to hear the same information many times before they can fully comprehend the problems they will encounter.

The concept of management rather than cure forms the basis of intervention. Long term aggressive therapy programs cause social isolation and delay normal psychological development. Integrate therapy programs into summer camps [B,C], home activities and school. Let the child live like a child, as close to normal as possible. Do not raise false hopes. Successful rehabilitation includes the prevention of additional problems, reduction of disability and community integration. Rehabilitation is successful if the child is a happy child and if the parents are well adjusted, happy people [D].

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Bracing

Braces are devices which hold the extremities in a stable position. The goals of bracing [A] are to increase function, prevent deformity, keep the joint in the functional position, stabilize the trunk and extremities, facilitate selective motor control, decrease spasticity and protect the extremity from injury in the postoperative phase. Their design and use requires an understanding of human biomechanics. Indications differ according to the age, selective motor control level, type of deformity and functional prognosis of the child.

The physician writes the prescription and the orthotist manufactures the brace. A close communication is necessary between them. Braces are custom-made from a plaster model of the child's extremity and fabricated with plastic materials. Evaluate the child for the first time together if possible. When the brace is ready, teach the child and the family how to put it on, take it off and how to look after it together with the therapist and the orthotist. Consider any modifications for better fit during this period. Evaluate the child once again after he begins to use the brace.

The brace should be simple, light but strong [B]. It should be easy to use. Most importantly it should provide and increase functional independence. The child will easily accept and use a brace which carries the above characteristics. Children will resist braces if the brace interferes with function. Do not insist if the child does not want to use the brace. The child quickly outgrows the brace in the growth spurt period. Indications change as the child's functional status changes. Evaluate the child at 3-6 month intervals and renew all braces regularly as the child grows.

Lower extremity bracing

Orthotics are usually named according to the body parts they cover [C]. Various kinds of ankle foot orthoses called AFOs are the most common braces used in CP [D]. Static braces immobilize the joint while flexible or hinged ones use body weight to stretch the muscles of the leg and ankle. AFOs provide appropriate contact with the ground during stance and foot clearance during swing. Knee immobilizing splints and hip abduction splints are prescribed both for nonambulatory and ambulatory children.

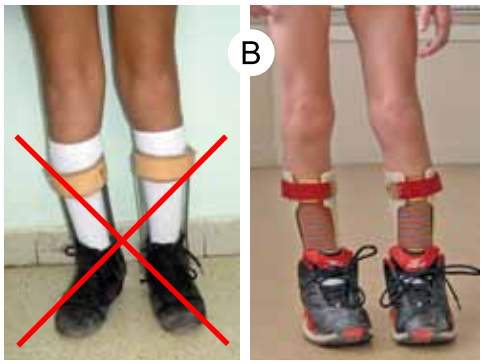
Compliance with night splints to prevent deformity is low. KAFOs work in children who use them. AFOs are not very useful as night splints because they do not prevent knee flexion.

Ankle foot orthoses (AFO)

The AFO [E] is the basic orthosis in CP and is a crucial piece of equipment for many children with spastic diplegia. The main function of the AFO is to maintain the foot in a plantigrade position [F]. This provides a stable base of support that facilitates function and also reduces tone in the stance phase of gait. The AFO supports the foot and prevents drop foot during swing phase. When worn at night, a rigid AFO may prevent contracture. AFOs provide a more energy efficient gait but do not prevent foot deformities such as pes valgus, equinus or varus.

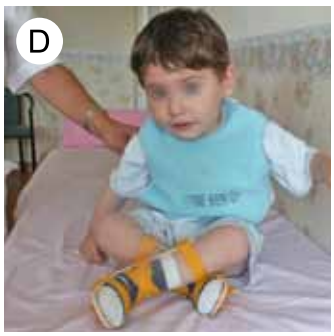
It is better to use the AFOs part time in most children. They may cause sensory deprivation and muscle atrophy if used continuously. Adolescents generally outgrow their braces and adults do not comply with them.

A	Goals of brace prescription
	Increase function
	Prevent deformity
	Keep joint in a functional position
	Stabilize the trunk and extremities
	Facilitate selective motor control
	Decrease spasticity
	Protect extremity from injury in the postoperative phase



The evolution of braces in CP: From metal and leather to plastic and carbon, with better understanding of the biomechanics; from KAFOs to AFOs with ankle control. Orthopaedic shoes, KAFOs and calipers have largely been abandoned. They are cumbersome, have very limited mechanical advantages, are very difficult to done on and off and in many cases they hide the deformity rather than correct it.

Braces in CP	C
Ankle foot orthoses: AFOs	
Knee-ankle foot orthoses: Plastic KAFOs and knee immobilizers	
Hip abduction orthoses	
Thoracolumbosacral orthoses: TLSOs	
Supramalleolar orthoses: SMOs	
Foot orthoses: FOs	
Hand splints	

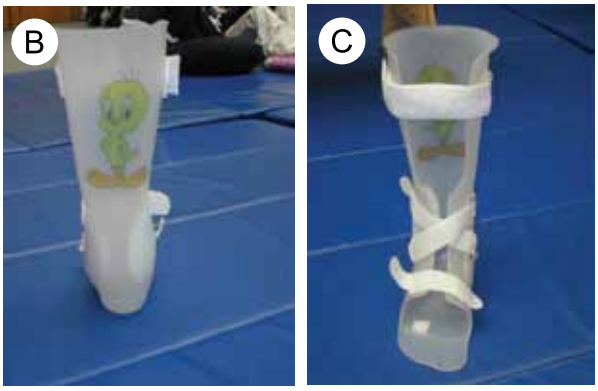


Children with CP do not benefit from and cannot tolerate extensive bracing such as total body braces, HKAFOs with pelvic bands and KAFOs. AFOs and their variations are generally sufficient to increase function, therefore an AFO is the basic brace in CP.

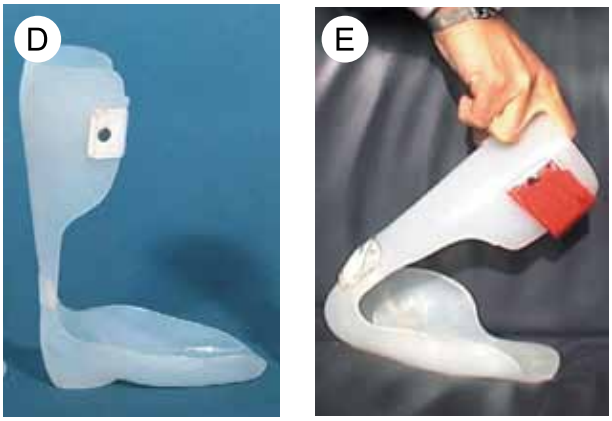
D: Courtesy of M. Sussman

Functions of the AFO		F
Main function	Keep the foot in a plantigrade position	
Stance phase	Stable base of support	
Swing phase	Prevent drop foot	
At night	Prevent contracture	

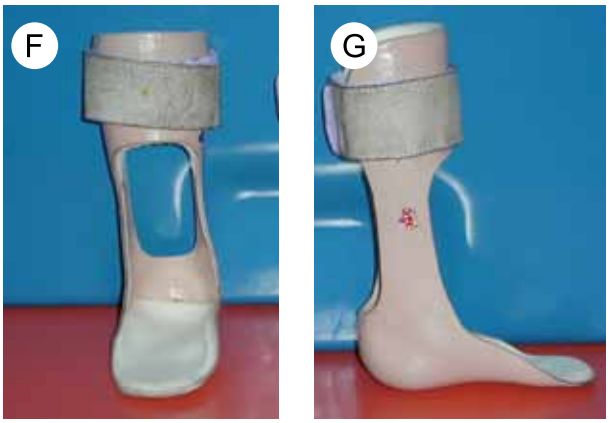
A	Types of AFO
	Solid AFO
	Posterior leafspring AFO (PLSO)
	Ground Reaction AFO (GRAFO)
	Antirecurvatum AFO
	Hinged AFO
	Hinged GRAFO
	Hinged antirecurvatum AFO



Solid AFO as seen from posterior and anterior



Posterior leaf spring AFO (PLSO) in neutral position and under load



GRAFO as seen from posterior, lateral and anterior. The characteristic of a GRAFO is the tibia restraint in front. This restraint prevents the tibia from moving forward as the person starts to put his weight on his extremity in stance (the second rocker phase of stance) It prevents excessive ankle dorsiflexion and crouch gait.

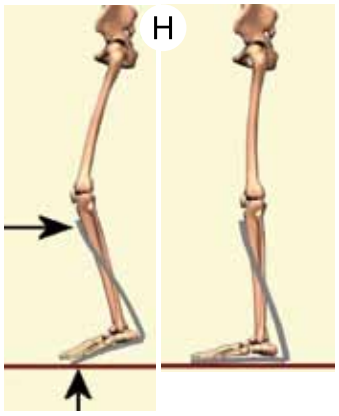
There are various types of the AFO [A].

Solid AFO The solid or rigid AFO [B, C] allows no ankle motion, covers the back of the leg completely and extends from just below the fibular head to metatarsal heads. Raise the sides for better varus-valgus control. The solid AFO enables heel strike in the stance phase and toe clearance in the swing phase. It can improve knee stability in ambulatory children. It also provides control of varus/valgus deformity. Advise solid AFOs to prevent contractures and to provide ankle stability in the standing frame in nonambulatory children. Because they are more comfortable compared to a short leg cast, consider using them in the early postoperative period for protection of the operated extremity.

Posterior leaf spring AFO A PLSO is a rigid AFO trimmed aggressively posterolaterally and posteromedially at the supramalleolar area [D]. This provides flexibility at the ankle [E] and allows passive ankle dorsiflexion during the stance phase. A PLSO provides smoother knee-ankle motion during walking while preventing excessive ankle dorsiflexion, particularly in larger children who have the strength to deform the material. However it also increases knee flexion in stance. Varus-valgus control is also poor because it is repeatedly deformed during weight bearing. The brace breaks when it is repeatedly deformed. These AFOs are frequently renewed because of material failure. A PLSO is an ideal choice in mild spastic equinus. Do not use in patients who have crouch gait and pes valgus.

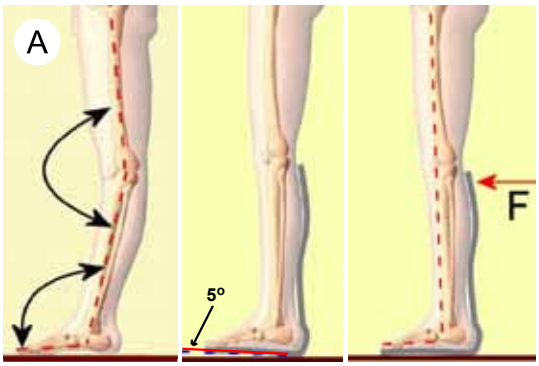
GRAFO or FRO (Ground reaction or floor reaction AFO) This AFO is made with a solid ankle at neutral [F]. The upper portion wraps around the anterior part of the tibia proximally with a solid front over the tibia. The posterior opening extends to the malleoli level. The rigid front starts just below the tuberositas tibia with a band at the back to create a three point pressure distribution and provide strong ground reaction support for patients with weak triceps surae [G]. The foot plate extends to the toes. The ankle may be set in slight plantar flexion of 2-3^{o} if more corrective force at the knee is necessary. Use the GRAFO in patients with quadriceps weakness or crouch gait. It is an excellent brace for patients with weak triceps surae following hamstring lengthening [H]. Use an anterior strap in children below 15 kg. Above that, use a rigid GRAFO if the foot alignment is poor and a hinged GRAFO if it is satisfactory. The benefit depends also on the work quality of the orthotist.

Children with static or dynamic knee flexion contractures do not tolerate the GRAFO. Surgically release the knee flexion contracture before prescribing the GRAFO.



Mechanism of action of the GRAFO: By pushing the tibia back, the GRAFO prevents passive ankle dorsiflexion in stance. When the tibia does not come forward, the femur rolls over the tibia and the knee extends.

Anti-recurvatum AFO This special AFO is molded in slight dorsiflexion or has the heel built up slightly to push the tibia forward to prevent hyperextension during stance phase [A]. Consider prescribing this AFO for the treatment of genu recurvatum in hemiplegic or diplegic children. Anti-recurvatum AFOs may be solid or hinged depending on the child's tolerance.



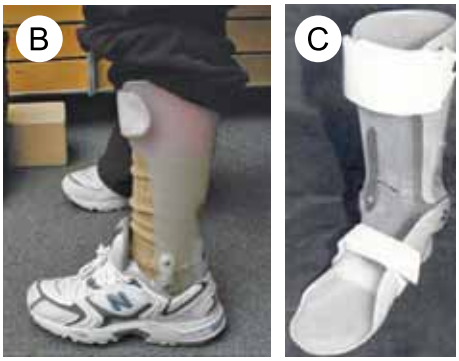
The mechanism of antirecurvatum AFOs: The AFO is built in 5° dorsiflexion. Therefore initial contact occurs with the ankle in dorsiflexion. Equinus is prevented. The back of the AFO pushes the tibia forward and the ground reaction force vector slides behind the knee joint creating a flexion moment at the knee,

Hinged AFO

Hinged AFOs have a mechanical ankle joint preventing plantar flexion, but allowing relatively full dorsiflexion during the stance phase of gait [B,C]. They provide a more normal gait because they permit dorsiflexion in stance, thus making it easier to walk on uneven surfaces and stairs. This is the best AFO for most ambulatory patients. Adjust the plantar flexion stop in 3-7º dorsiflexion [D] to control knee hyperextension in stance in children with genu recurvatum.

The hinged AFO is contraindicated in children who do not have passive dorsiflexion of the ankle because it may force the midfoot joints into dorsiflexion and cause midfoot break deformity. Knee flexion contractures and triceps weakness are other contraindications where a hinged AFO may increase crouch gait [E].

Children with quadriplegia and diplegia benefit more from solid AFO's that control pes valgus, while posterior leaf spring AFO's or hinged AFO's are more suitable for kids with hemi or monopleagia, especially for those who have active dorsiflexion.



The AFO may be fitted with a hinge that allows 10° passive dorsiflexion while preventing plantar flexion. This creates a more natural gait but the hinges may be an obstacle to wearing shoes.

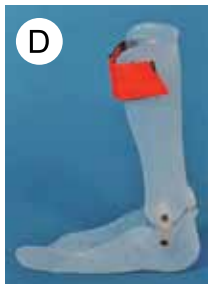
It unclear whether AFOs can maintain or increase muscle length and hence prevent or reduce deformity.

Knee orthoses

Knee orthoses are used as resting splints in the early postoperative period and during therapeutic ambulation. There are two types of knee orthoses, the knee immobiliser and the plastic knee-ankle-foot-orthosis (KAFO). The use of such splints protects the knee joint, prevents recurrence after multilevel lengthening and enables a safer start to weight bearing and ambulation after surgery.

Knee immobilisers

Knee immobilisers are made of soft elastic material and holds only the knee joint in extension, leaving the ankle joint free [F]. Consider using them in the early postoperative period after hamstring surgery and rectus transfers.

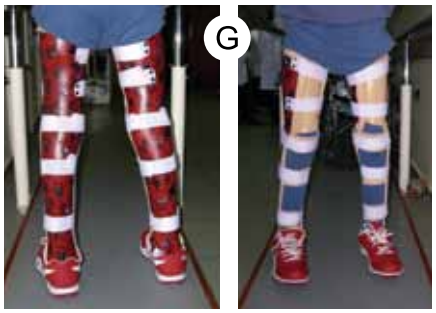


Hinged antirecurvatum AFO

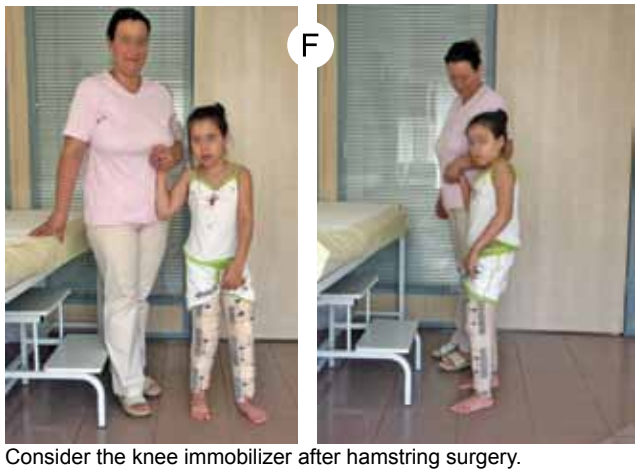
Hinged GRAFO

Plastic KAFOs

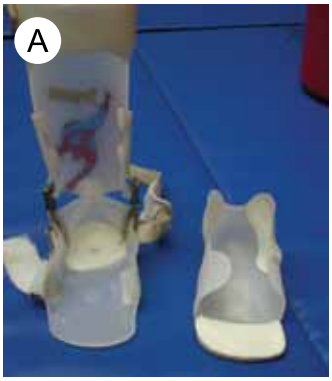
Plastic resting KAFOs extend from below the hips to the toes and stabilize the ankle joint as well as the knee [G]. They are more rigid and provide better support to the ankle and the knee in the early postoperative phase.



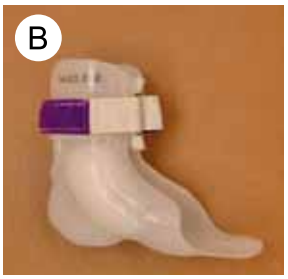
Use the plastic KAFO at night and in the early postoperative period after multilevel surgery to protect the extremity while allowing early mobilization.



Consider the knee immobilizer after hamstring surgery.



SMOs can be used inside hinged AFOs to provide better valgus -varus control while allowing ankle dorsiflexion.



Supramalleoler orthosis (SMO)

UCBL

The spinal brace becomes part of the total body involved child.
Courtesy of J. Batzdorff

E	The indication for spinal braces
	To slow the progression of deformity to delay surgery and allow skeletal growth
	To assist sitting balance
	To protect the surgical site from excessive loading after surgery

Knee-ankle-foot orthoses with metal uprights and hinged joints (KAFOs) were developed and used extensively in the 1950s and 60s for children with poliomyelitis. Though KAFOs are still used for ambulation in poliomyelitis and myelomeningocele where there is a need to lock the knee joint, they are not useful for the child with CP because they disturb the gait pattern by locking the knee in extension in the swing phase. Donning the KAFOs on and off takes a lot of time and they are difficult to wear. For these reasons, KAFOs for functional ambulation have disappeared from use in children with CP. Use anti recurvatum AFOs or GRAFOs for knee problems in ambulatory children.

Foot orthoses (FO)

Foot orthotics do not prevent deformity. They provide a better contact of the sole of the foot with the ground.

Supramalleoler orthosis (SMO) Extends to just above the malleoli and to the toes [A,B]. Consider in mild dynamic equinus, varus and valgus instability.

University of California Biomechanics Laboratory Orthosis (UCBL) Medial side is higher than the lateral, holds the calcaneus more firmly, supports the longitudinal arch [C]. Prescribe in hind and midfoot instability.

Heel cup Holds the calcaneus and the surrounding soft tissue, ends laterally underneath (trim lines are below) the malleoli and proximally ends at the metatarsals. Use in cases of mild subtalar instability causing varus or valgus deformity.

Hip abduction orthoses

Consider using hip abduction orthoses in children with hip adductor tightness to protect hip range of motion and prevent the development of subluxation. It is easier and cheaper to use a simple abduction pillow. Use mainly at night or during periods of rest. There is no scientific evidence to support the belief that they prevent subluxation. One clear indication for hip abduction orthoses is the early period after adductor lengthtening.

Spinal orthoses

There are various types of braces used for spinal deformity [D]. None of them alter the natural history of scoliosis in children with CP. Do not aim to stop the progression of scoliosis by prescribing a brace. Contrary to idiopathic scoliosis, the deformity continues to progress even after skeletal maturity in CP. Therefore, most children with scoliosis need spinal surgery to establish and maintain sitting balance in the long run. Prescribe a brace for the time period until surgery to enable the child to grow as much as possible. An important indication for using a brace in a spinal deformity is to provide better sitting balance [E]. A thoracolumbosacral brace helps the child sit better during the growth spurt period when spinal deformity becomes apparent, progresses fast and the child outgrows custom molded seating devices quickly. Children who are not candidates for surgery for different reasons may use spinal braces instead of seating devices for better sitting. Patients with mild and early scoliosis tolerate brace without difficulty. The brace should not be too difficult for the child to put on and take off should not compress the chest too tight and should be properly ventilated for comfortable use.

Upper extremity bracing

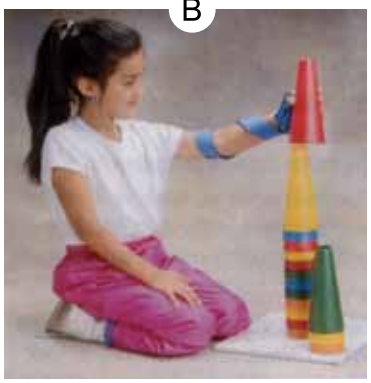
The indications of bracing in the shoulder and elbow are very limited. There is no good evidence to support claims that wrist hand orthoses prevent or correct deformities and improve manual ability over time.

An example of a resting splint [A] is a thermoplastic resting elbow, wrist and hand splint which keeps the wrist in 10° extension, the metacarpophalangeal joints in 60° flexion and the interphalangeal joints in extension. This type of splint is used at night and during periods of inactivity with the hope of preventing deformity.

An example of a functional splint is an opponens splint [B] to bring the thumb out of the palm of the hand, allowing for better grasp. This type of splint is used in every day activities. However, it is still unknown whether a thumb abduction orthosis improves use and manual function of the affected hand in children with hemiplegia.

Hand orthoses may inhibit the active use of the extremity. They also effect sensation of the hand in a negative way. Use them only in the therapy setting or at school and take off during other times in the day.

Resting hand splint Copyright Smith & Nephew



Functional hand splint
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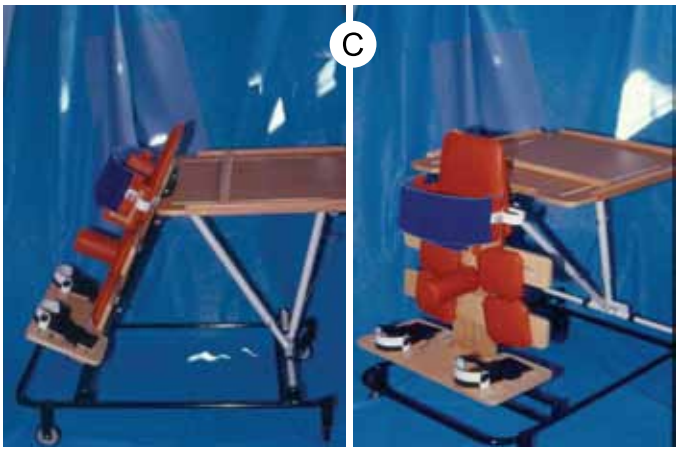
Mobility aids
Standers
Walkers
Crutches
Canes

A

Advantages of mobility aids
Develop balance
Decrease energy expenditure
Decrease loads on joints
Improve posture

B



A prone stander supports the body from the anterior. Tilting standers enable the child to slowly get accustomed to standing position.
Do not use prone standers if the child cannot hold his head. Children with increased extensor spasticity tend to arch their back and they are not comfortable in prone standers. Courtesy G. Koloyan

D

Benefits of standers
Support erect posture
Enable weight bearing
Stretch muscles to prevent contractures
Decrease muscle tone
Improve head and trunk control

E

Consider these factors when selecting a stander
Head and trunk control
Abnormal postures and muscle tone
Growth
Potential for mobilisation

Supine standers are easier to use in large children and children who do not have head support.

Mobility Aids, Wheeled Mobility & Assistive Devices

A child with CP needs to move around, to explore his surroundings and to interact with his peers so that his mental, social and psychological skills develop to the fullest. A variety of mobility aids and wheelchairs provide differing degrees of mobility to these children [A,B]. Transfer aids such as lift systems assist the caregiver when performing transfers. Passive standing devices called standers [C] allow the child to get accustomed to standing erect and provide therapeutic standing. Some ambulatory children have to use gait aids as well as braces for efficient and safe ambulation. These gait aids are walkers, crutches and canes. They are mainly used to assist with balance, not for weight bearing.

Gait aids help develop balance. The child receives sensory information regarding the position of the body and space by holding onto the walker, crutch or cane. Gait aids decrease energy expenditure, decrease the loads on the joints, improve posture and pain in addition to improving balance. Children who do not have ambulation potential need to use wheelchairs for moving around. Wheelchairs must be properly chosen and fitted with seating aids, cushions and other positioning components.

Transfer aids

Different types of lifts provide safe and easy transfer of the older and heavy handicapped children. These are designed to help the caregiver transfer the child without trunk and upper extremity control into wheelchairs, toilet and baths. Some families prefer lift systems that have slings for the child to sit in.

Transfer boards are simple rotatory or sliding devices to move the child from bed to the wheelchair. The child sits on a round board that rotates 360°. When the board rotates to position, the child slides from the bed to the wheelchair.

Grab bars and rails may be placed around and over the bed to improve bed mobility.

Standers

Standing in an erect posture contributes to the child with CP in many ways [D]. It establishes the sense of verticality, helps develop better eye contact, improves communication and balance reactions. The pulmonary, cardiovascular, gastrointestinal and urological system functions all improve by standing. Passive standers support the child in the erect posture and enable weight bearing on the lower extremities, stretch the muscles and may prevent contractures, decrease muscle tone, improve head and trunk control. There are supine standers, prone standers and the parapodium [E].

Prone frames

Prone frames support the body and the chest from the front. This position stretches the hip flexors, providing knee extension and ankle dorsiflexion. Lateral body supports, hip guides, abductor blocks, knee blocks and shoe holders support the body and the extremities. If the head control is not satisfactory, the child may use a chin support. The angle between the ground and the frame can be adjusted to stimulate head and body control against gravity. The tray in front helps the child to put his weight on the upper extremities and also use them actively. Prone frames stimulate the child to actively use the extensor muscles of the back. Do not use prone frames in children with poor head control and increased extensor spasticity. Choose supine frames in such children.

Supine frames

Supine standers support the child from the back [F on previous page]. Lateral supports, knee pads abduction-adduction hip supports and head rests help maintain standing. Supine standers are better for children who do not have head control and who need to work on upper extremity skills. Prefer to use them when there is extensor muscle spasticity. Supine standers are easier to use in large children. Begin to use standers as early as possible to adapt the child to erect posture. Encourage the nonambulatory child to spend some of the day in the standing frame and work on upper extremity function in the meantime. Gradually increase the time in the frame from 15 minutes to a couple of hours at different hours during the day. The child with walking potential learns to stand in the stander and then gradually progresses to training in parallel bars.

Courtesy of M. Sussman

Gait trainer

The gait trainer is a metal frame with metal uprights that support the trunk and arms. Slings or bicycle type seats are attached to keep the child erect. It provides significant trunk and pelvis support and can help teach the child a reciprocal gait pattern. Consider using the gait trainer to prepare the child for walking. As the child gets used to walking in a reciprocal manner in the trainer, he progresses to walking with a simpler assistive device such as a walker or crutches. The gait trainer is also useful in to provide therapeutic ambulation at home in total body involved children.

Gait aids

Walkers

All children's walkers should be built from ultralight durable aluminium and supplied with wheels to minimise energy expenditure. Swivel wheels, forearm attachments, hip guides, hand brakes, baskets and seats are added to the walkers if necessary. Walkers provide the greatest support during gait but they pose certain difficulties during stair climbing, among crowds and within narrow corridors.

There are two types of walkers for pediatric use. The anterior open (reverse) [A, B] walker is also called a postural control walker. Use the reverse wheeled walker in the majority of children. It provides the best gait pattern and is less energy consuming [A on next page]. Standard forward walkers lead to increased weight bearing on the walker and increased hip flexion during gait. Choose them only in cases where extensor spasticity predominates [C].

Do not use standard walkers in children with CP except if there is severe extensor spasticity. The reverse walker is the primary choice because it provides an energy saving gait pattern and prevents hip and knee flexion.

Canes, crutches and gait poles

Walking aids are usually prescribed for balance problems. Slowly push the standing child from the side and then from the front and back. Watch for signs of disturbed balance. Canes or gait poles are necessary if the child does not have sufficient lateral balance. Quadriped canes are the next usual step as the balance improves following walker use. Instruct the child to use the quadriped canes lateral to the body rather than out in front. Try to switch from posterior walker to forearm crutches in adolescents. Gait poles [D] or sticks provide sensory input for gait and facilitate a normal gait pattern, but sometimes are not cosmetically acceptable to patients. Avoid forearm crutches, as children tend to lean forward into these and develop hip flexion contracture [E]. Forearm crutches also lead to the child to bear the body weight on the upper extremities, leading to a pattern of walking on all fours. Use forearm crutches only in children who need an assistive device for weight bearing as well as balance.

Gait poles are sticks that the child holds near his body to maintain balance. They provide a straighter posture compared to crutches.
Courtesy of M. Sussman

The child using crutches tends to go into a flexion pattern which increases the energy consumption of walking.

Children with spastic diplegia have problems with posterior balance and tend to fall backwards. In this case, prescribe a walker. The child who walks very slowly in high guard position and falls backwards after a few steps benefits from a reverse walker as an aid to improve his balance.

Collapsible and lightweight manual wheelchairs can be used outdoors for mobility.

Most total body involved children prefer to use power wheelchairs because involvement of the upper extremities makes them unable to use manual wheelchairs.

D **Wheelchair use in terms of function**

Independent	Independent in sitting and rising from a wheelchair.	Manual or power wheelchair
Needs help in transfers	Somebody is necessary to help to sit in and rise from a wheelchair.	Manual or power wheelchair
Dependent	The child is carried when sitting in and standing up from a wheelchair.	Strollers, wheelchairs which tilt backwards, or wheelchairs with reclining backs

Wheelchairs

Encourage wheeled mobility in all children who have poor potential for walking. Strollers and wheelchairs are options for wheeled mobility. Use strollers, wheelchairs which tilt backwards, or wheelchairs with reclining backs in children who are totally dependent and do have any potential for independent mobility. They provide caregiver relief in transport and ease of care.

A wheelchair is a mobility as well as a seating device (positioning device) in children with severe motor dysfunction, poor sitting balance and no functional ambulation [B]. Independent mobility can be achieved with manual or power wheelchairs in children who have adequate cognitive and motor function. Independent wheeled mobility allows the child to explore his surroundings, contributes to his mental improvement, socialization and self esteem.

Prefer motorized wheelchairs in severely involved children who have upper extremity dysfunction [C]. Motorized wheelchairs have a great positive impact on the life of the severely impaired child and his family. Consider prescribing them as early as four-five years of age and enable the child to move around independently to explore his surroundings and to take part in family life without spending too much energy.

Some severely involved spastic and athetoid children spend excessive energy while trying to walk with walkers and crutches. Motorized wheelchairs preserve energy and improve the level of social and educational function. Even though there is an argument that early use of the motorized wheelchair causes laziness and decreases cardiovascular capacity it is obvious that the beneficial effects of early independent mobilization with less energy expenditure far outweigh the risks.

Choose an age-appropriate mobilization device and teach the child how to use it [D].

Factors in wheelchair prescription	
Foot rests	Long enough for the shoe to fit inside Hold the feet in neutral position Able to swing out of the way Velcro bands for restraint if feet control is poor
Seating	Height: feet placed firmly against the foot rests ankle in neutral, hips & knees in 90° flexion Depth: support both thighs not compress the poplitea Width: wide enough to relieve the trochanter prevent the pelvis from slipping sideways Firmness: to limit of tolerability for maximum stability prevent pressure sores over the bony prominences
Back	Height: the middle of the scapula Width: accommodate the trunk Supportive peds inside Semirigid to prevent kyphosis Custom molded body braces for scoliosis Reclining
Portability	Light to fit inside an automobile
Propulsion	Sufficient upper extremity function: self propelled dysfunctional upper extremity: caregiver propelled

A

There are many factors to consider when prescribing a wheelchair [A]. Always make sure that the chair is comfortable for the child. Do not use the wheelchair to stretch the spastic muscles, because this will be too uncomfortable for the child [B,C].

Seating systems

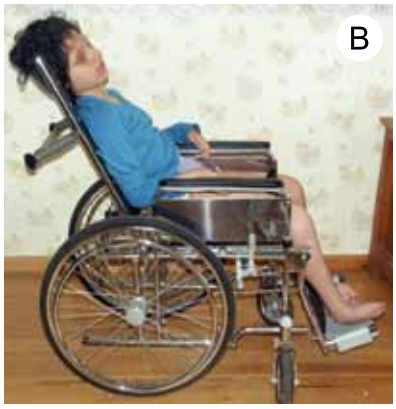
Seating systems provide support and stability, prevent postural deformity and enable the use of upper extremity in the severely impaired child without trunk control. It is presumed that good seating promotes normal development by maintaining of postural alignment and reducing undesirable tone and reflexes; enhances postural stabilisation of the trunk as a prerequisite to functional performance of upper limb; helps to prevent or delay development of deformity or muscle contractures; optimises the child's position for feeding, respiratory and digestive function; assists exploration of the child's environment; and improves head control which is essential for orientation and socialisation and to help the child to develop cognitive and communication skills. In short, a seating system is usually chosen with the aim to improve posture and enhance function.

The seat should be combined with hip abduction, foot rests, hip belt, and a cut-out tray as appropriate to enable the optimum sitting posture determined individually for each person [D].

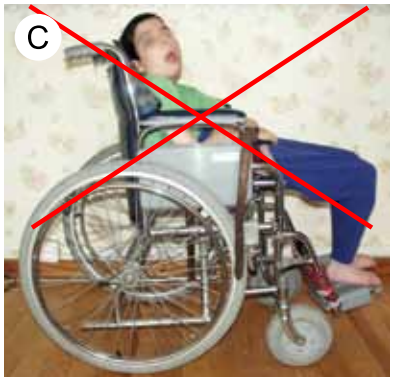
Seating devices have various components. They can be made both for the back and for the seat. They are either linear, contoured or custom-molded [E].

Linear systems

Basic material for linear systems consists of wood for the base, foam for comfort and pressure relief and a cover. They can also have abductor or adductor supports at the sides. Linear systems compensate for the child's growth and accommodate to the size. They are not comfortable and insufficient to relieve pressure over the bony prominences.



The wheelchair must be very comfortable. The back of the chair should have a slight inclination of 110° to the ground in children with poor trunk control. This position also prevents excessive stress on the spastic hamstring muscles. Posterior tilting of the seat does not prevent deformity; on the contrary, upright/anteriorly tilted sitting positions appear to be more functional for seeing, eating and using the upper limbs.



If the wheelchair is too deep, the spastic hamstrings pull and the patient slides out of the chair. This will create the false impression that the chair is too small for the patient. Always check to see whether the pelvis is properly aligned in the chair.

Hip and spine alignment depend upon the pelvis which should be horizontal in the wheelchair. Seating systems aim to keep the pelvis this way. Add neck and arm rests, knee pads, foot supports and adductor pillows if necessary.

E

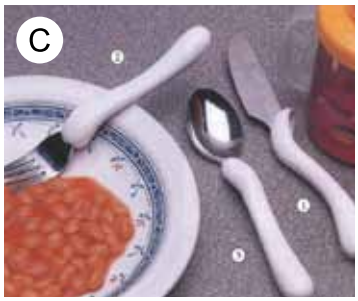
Seating systems			
	Linear	*Contoured*	*Custom-molded*
Trunk support	Good	Better	Best
Comfort	Good	Better	Best
Accommodate to size	Yes	No	No
Expense	Less	More	Most

A good cushion
Provides enough support over the bony prominences
Creates a stable sitting base
Durable
Light
Usable in hot seasons

A

B

Attach a headrest to the wheelchair or stroller to provide head support.
© Meyra

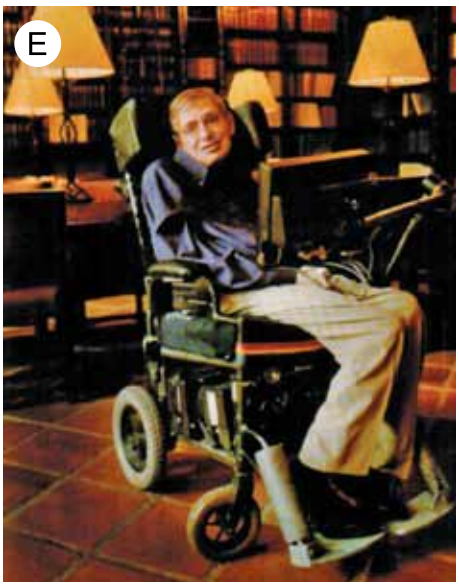


C

Add foam tubing to the handle to build up the grip or bend the utensil to improve the angle to bring the food to the mouth easily.

D

An advanced communication board produces sounds when the picture is touched.
C,D: © Nottingham Rehab Supplies



E

The famous scientist Stephen Hawking wrote his world famous books using such an alternative communication device.
From: S. Hawking: The Universe in a Nutshell. Photo: S. Cohen

Contoured systems

Contoured systems are better aligned to the body. They do not accommodate to growth so one must renew the system frequently if the child is growing fast. This increases the cost of treatment .

Custom-molded systems

Custom-molded seating systems provide the most trunk support. They enable sitting in children who have complex deformities. They are expensive, need to be replaced as the child grows, and limit the child's movements in the seating device.

Cushions and positioning components

Various cushions are manufactured to provide an even distribution of body weight and to prevent pressure sores [A]. They contain either foam, water, air or a gel-like substance. Light cushions are advised if the child is independent in transfers.

Adding certain positioning pieces to the seating system provides better trunk alignment in the wheelchair dependent child. Side supports keep the body in the center while chest belts support the front. A pelvic band at 45° to the seating surface positions the pelvis. Use abduction pillows and wedges to prevent excessive adduction of the hips. Footrests are helpful to position the feet correctly.

The position of the head is important for many reasons including visual perception, control of muscle tone, feeding and swallowing. Posterior and lateral head rests provide support and increase transport safety in children who do not have head control because of low muscle tone [B].

A child sitting in a properly fitted wheelchair with the right seating and positioning devices has more opportunity to explore and experience the world. Social integration and involvement in school activities increases greatly.

Assistive aids

There are a variety of assistive devices used in children with CP to gain function. These devices aim to decrease the caregiver's burden and to increase the child's independence in activities of daily living, communication, education, recreation and vocation.

Feeding aids

Various knives and forks have been devised to enable independent feeding [C]. The shape, thickness and angle of these knives, forks and spoons are modified according to the child's joint range of motion, strength and coordination. These kitchen utensils may be bought or modified from standard knives and forks at home. Mechanical and electronic feeding devices have also been invented for children who do not have sufficient hand control but they are very expensive and not available worldwide.

Aids for communication

Communication is among the most important priorities of children with CP. Communication impairment has two components, dysarthria and dysphasia. Problems of speech and articulation are called dysarthria and problems of language are named dysphasia. Dysarthria occurs because of involvement of the oropharyngeal or laryngeal muscles. Dysphasia occurs because of mental problems secondary to global developmental delays or because of a lesion in the language centers of the brain.

Various devices exist to improve both speech and language impairments and increase communication. They range from very simple picture sets of symbols to high technology equipment such as computerized systems. The simplest is a communication symbol set used to understand the child's wishes. It can be made at home from simple pictures. The child learns the meanings of these symbols in activities of daily living. Speech therapists teach the child how to express his thoughts, needs and feelings using communication boards, notebooks and devices producing simple every day talk. Communication boards are a set of symbols and pictures that the child sees and knows from everyday life [D on previous page]. He simply points at the picture or nods when the picture is pointed at. More complex systems produce sounds when the picture is pushed.

Computerized systems [E on previous page] developed after 1980s produce age and gender appropriate speech in different languages. A personal computer or portable notebook computer working with mouse, keyboard, joystick, eye gaze, touchscreen or breath supplied with the appropriate software is necessary for speech production. The speech impaired child can communicate with his family and friends in this manner but the language impaired child will still need picture boards and symbol sets.

A computer is an asset for the child who cannot speak but can write. Dyskinetic and total body involved children who cannot speak but have sufficient mental function can use it with an appropriate mouse or trackball to write and express themselves. All children who have problems with fine motor control benefit from having a computer at school for education.

Recreational equipment

Games are the primary means of any child to discover the world and learn. The child with CP needs to join the community life, playing games with peers and friends. There are many simple and relatively cheap options to increase the child's opportunity to play. The three-wheeled bike can be modified for the disabled child with hand propulsion, wide seats, seat belts, trunk supports and chest straps [A]. Children with trunk extensor spasticity can pedal special bicycles in the upright position [B]. Battery powered cars can easily be adapted with joysticks or special switches.

Special tricycle with modified handles, head rest and restraints for the body.

Two small rear-wheels are added to a standard bicycle for better balance.

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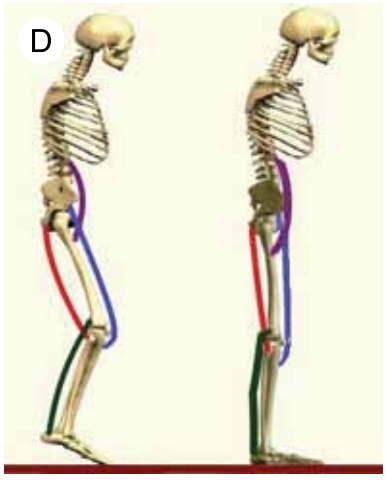
Orthopaedic surgery is the treatment of choice in children with musculoskeletal problems not amenable by conservative means.

B

Goals of orthopaedic surgery		
	Goal	*Operate to achieve*
Walking potential	Functional ambulation	Good hip and knee extension Stable hips Plantigrade stable feet
No walking potential	Sitting balance	Straight spine Horizontal pelvis Stable hips

The effects of a bad and a good posture are easily distinguished in this diplegic child before and after surgical treatment. A good posture is essential for efficient ambulation.



Orthopaedic surgery effectively corrects posture by achieving balance between muscle groups and eliminating bony deformities.

Orthopaedic Surgery

Orthopaedic surgery is widely used in the management of children with CP to prevent or correct certain musculoskeletal problems such as muscle shortening and bony deformities [A]. The goal of orthopaedic surgery in a child with walking potential is to improve functional ambulation. For non-ambulatory children, the goals of orthopaedic surgery are to facilitate sitting, improve hygiene and prevent pain [B].

Aim to obtain plantigrade stable feet, stable hips, good hip and knee extension in the ambulatory child [C,D]. Make sure that the hips are stable, the pelvis is horizontal and the spine is straight in the nonambulatory child. Be aware of and prevent deformities caused by muscle imbalance which in later life will cause pain and loss of function. Surgical correction of deformities improves body and self image and provides psychological satisfaction.

Surgery cannot be the single solution to the wide variety of problems caused by CP. It is only a momentary pause in the long journey of CP management. The need for rehabilitation measures such as bracing, physiotherapy, sports and antispastic medication still remains after surgery.

Preparing for surgery

Family cooperation is essential for the success of treatment. Set realistic goals after a thorough evaluation of the expectations and limitations of the family. Discuss the expected surgical outcome with them to the extent of their understanding [E]. Reconsider the treatment plan if the expectations of the family are not consistent with the aims of surgery. Family counselling can be helpful to overcome the communication barriers and to prevent disappointments. Sometimes a member of the treatment team may have a better dialogue with the family and can help develop communication and mutual understanding. The team must support each other and each team member must provide the family with the same information. The family will then trust and put their confidence in treatment.

All deformities of the musculoskeletal system can be corrected. However, there are times when the correction of deformity does not necessarily result in functional gain. The patient may look better after surgery but does not necessarily function better. There are certain patients in whom surgical procedures cause more complications than benefits. The multitude of physical problems and the various treatment procedures create a psychological burden in the child and the family. Surgery is an additional cause of major stress in this patient group. Be realistic and define what

E

What to tell the family before soft tissue surgery
We aim to decrease the tight muscles that hinders your child's ability to stand and walk
We will lengthen the muscles to help your child stand and walk better.
He will be able to hold his legs apart and sit upright.
However;
We are treating the consequences of the disease, not the disease. Your child will still have CP after the operation.
He will need a cast for few weeks and a brace for 3 - 6 months.
He may still have balance problems so he may still use his walker or crutches.
His muscles will be weaker, so he will need a lot of therapy.
It may take him several months to regain his preoperative abilities.

you hope to gain by surgery in each individual patient. Make sure that the operation will create considerable functional gains not obtainable by other means.

Improvements expected by surgical procedures

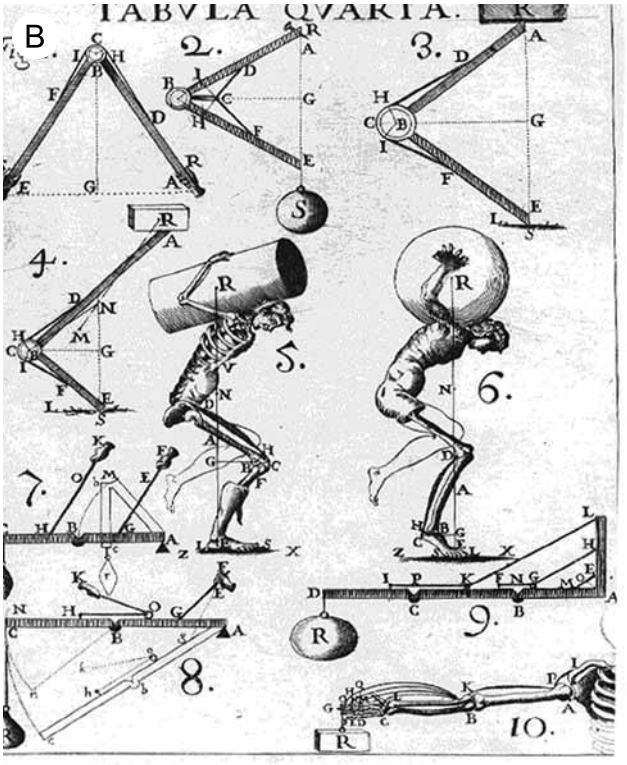
Orthopaedic surgery corrects some of the primary and secondary impairments in CP [A]. First, it reduces muscle tone because lengthening of the spastic muscles decreases the sensitivity of the stretch reflex. Balance is decreased immediately after surgery but improves in the long run because the patient has plantigrade stable feet which provide a better base of support. Muscles usually get weak after surgery but they respond well to strengthening exercises. Tendon transfers change the direction of deforming forces that create muscle imbalance. Tendon transfers may prevent deformity and allow the child to use his muscle strength more efficiently in this way. Decreasing spasticity of the antagonist muscles allows the agonists to function better. Improving balance by creating a stable base of support also helps movement. These factors indirectly improve selective motor control, however primitive reflexes do not change after surgery.

Most importantly, surgery corrects deformities of the spine and extremities that disturb sitting, standing and walking capacity. The human musculoskeletal system can be regarded as a system of multiple lever arms where joints act like fulcrum points, the bones are the lever arms and the muscles provide the necessary force [B]. This approach of the lever system provides a better understanding of most orthopaedic problems and especially the musculoskeletal problems of CP patients. When the joints are malaligned, the bones are malrotated or the muscles are weak, or do not pull in the desired direction, the lever system cannot perform efficiently. For example, a dislocated hip joint is a bad fulcrum and the forces acting through this fulcrum cannot perform efficient work. Another example related to CP is a weak tibialis anterior muscle that acts as a weak lever arm and cannot dorsiflex the foot to oppose the spastic gastrocnemius muscle [C].

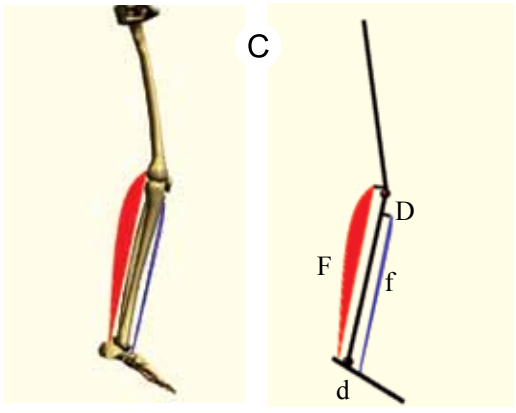
Tendon lengthening decreases the unopposed pull of spastic muscles and prevents skeletal contractures and deformities caused by this pull. Muscle balance determines posture in sitting and standing. Orthopedic surgery reestablishes this balance by lengthening and transfers to provide stable standing and sitting [D].

Impairment	Expectation after surgery
Primary impairments (due to the brain lesion)	
Muscle tone	Decreases
Balance	Decreases immediately, then improves
Strength	Decreases
Selectivity	May improve
Secondary Impairments (due to the primary impairments)	
Contractures & deformities	Corrected, may be prevented

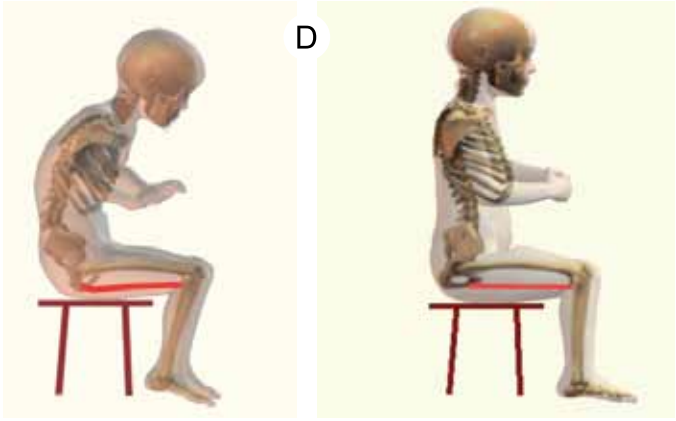
A



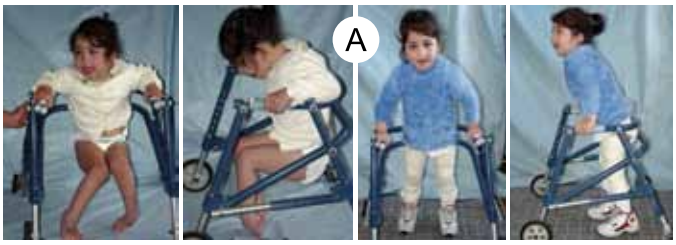
The human musculoskeletal system works as multiple levers as described by Borelli in the XVII. century. These levers do not function properly in CP. When the skeletal alignment is disturbed some of the force output that the lower extremity muscles produce becomes useless because they pull in the wrong direction. This is termed 'lever arm disease' by some experts.



The relatively weak tibialis anterior muscle (drawn as level arms and forces on the right) cannot create an efficient moment to counteract the pull of the spastic gastrocnemius muscle, the ankle remains in equinus.



The spastic hamstring muscles pull the pelvis posteriorly and cause posterior pelvic tilt. The child sits on the sacrum and lumbar kyphosis increases. Lengthening the hamstrings corrects this posture by allowing proper pelvic alignment.



Even severe contractures can be treated effectively with muscle lengthening. This is a 4 years old diplegic girl who had severe contractures before the operation and began ambulating with a walker postoperatively.
Courtesy G. Koloyan

Normal femur [B] is in slight internal rotation and the tibia compensates with mild external rotation. Persistent and increased femoral internal rotation in CP [C] causes intoeing. As the child grows, the tibia shows external rotation and the foot turns into valgus [D].

The muscles of the child with internally rotated femurs and externally rotated tibiae do not work in the plane of forward movement, muscle force is spent in the transverse plane. In the diagnosis of the torsional problems look to the patella when the child is walking. The patellae (red rings) are turned medially. Derotation osteotomy corrects the malalignment and enables the muscles to work in the correct plane.

Even severe contractures can be treated effectively with muscle lengthening [A].

Rotational osteotomies and arthrodeses that correct bony deformities help transfer the malaligned muscle force into the correct plane of movement, and make it easier for the child to walk [B-E]. Pelvic obliquity and painful hip dislocation can be prevented. Posture may be improved. The correction of joint alignment makes walking easier and the child may stop using coping mechanisms and adaptive responses he developed because of his contractures and deformities.

Timing of surgery

There are no absolute rules regarding time of surgery, only guidelines exist. These guidelines depend on the maturation level of the CNS, the walking potential of the child and the rate of deformity development. Use nonsurgical means to alleviate muscle tightness until the nervous system is mature. This occurs around the age of 4 to 6. At this age the physician can know more accurately what the muscle imbalance consists of, assess the functional prognosis of the child better and can make sure that no other abnormalities such as athetosis or dystonia are present. As a general rule, perform soft tissue procedures between ages 4 to 7, hand surgery between ages 6 to 12 and bony procedures after 8 years of age unless the rotational abnormality significantly deters their ability to walk. The exceptions to this rule that make early surgical intervention obligatory are progressive hip instability and early deformities and contractures interfering with function.

The age of surgical intervention needs to be tailorized for each patient even though certain age limits exist [F]. There are some children who may benefit from early surgery, and some children in whom musculoskeletal development continues above the age of 4 to 7 where surgery is best delayed. It is not the age but the needs of the patient which determine the timing and indication of surgery.

Delay upper extremity surgery to an age when the child's motor function can be clearly defined. Selective motor control is the key to hand function. It is appropriate to delay the procedure until selective control is established if the procedure requires the patient to selectively use certain muscles postoperatively. Perform upper extremity surgery between the ages of 6 to 12 when the child will cooperate easily with postoperative rehabilitation.

Patient selection

The skill of the orthopaedic surgeon lies partly in his ability to decide whether the patient will benefit from surgical intervention [A on next page]. Certain patients benefit a lot from orthopaedic surgery whereas others may get worse. Spastic diplegic and hemiplegic children improve more after

F

Timing of surgery	
Soft tissue procedures	Age 4-7
Hand surgery	Age 6-12
Bone procedures	Around puberty
Exceptions	
Progressive hip instability or severe femoral anteversion	
Early severe deformity interfering with function	

surgery compared to spastic total body involved, dyskinetic and mixed types. Fewer operations are performed and the gains are limited in dyskinetic cases.

Factors to consider in patient selection [B]

Neurological impairment

Functional gains depend on the extent of the lesion in the CNS. Orthopaedic surgery corrects deformity, balances the muscular forces across the joints and decreases spasticity but the patient can walk only if he has sufficient neurological function.

Selective motor control Orthopaedic surgery is usually performed to correct pes equinus and pes varus in hemiplegia and jump, scissoring and crouch gait in diplegia. In the child with total body involvement, spinal deformity and hip instability are treated with surgical methods. Results are best in children who have selective motor control [B]. Do not operate on dyskinetic cases.

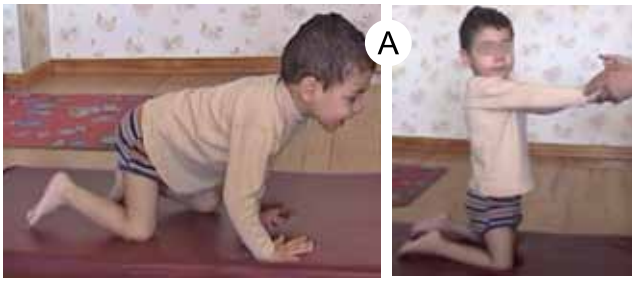
Balance has an important role in the functional status of the spastic child. However lack of balance is not a contraindication for an operation. Creating a stable base of support through surgery improves overall balance.

Cognitive function & visual impairment Children with cognitive problems need special evaluation [D]. Cognitive deficits and visual impairments are not by themselves contraindications for orthopaedic surgery and do not affect the surgical outcome unless they are very severe. The goal of surgery in severe mental retardation is to improve nursing care and relieve pain. Surgical correction of deformity may improve walking in a child with moderate cognitive deficit. Avoid surgical procedures requiring postoperative intensive physiotherapy or long term cast immobilisation in children with severe cognitive deficits.

Sensation is fundamental to hand function. Most children with upper extremity problems are not candidates for surgery because of sensory deficits. Consider hand surgery to gain function in the occasional spastic hemiplegic child who has good stereognosis. Perform surgery to improve hygiene and cosmesis in total body involved children.

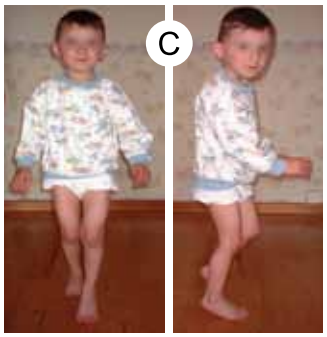
Apraxia Some children with CP have deficits in motor planning called apraxia. They are unable to move their body parts automatically through a sequence when carrying out a complicated action such as opening a door and walking out. This problem is difficult to diagnose and may be a reason why surgery does not improve function as much as expected in some cases.

Patients are grouped in terms of their functional capacity by the GMFCS [E].

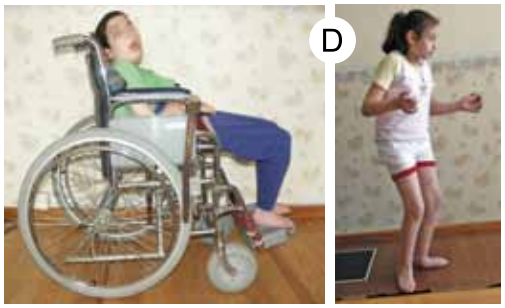


Put the child in a kneeling position to eliminate the effects of deformities. If the child can crawl on all fours, has balance and ability to move, then consider correcting the deformities for functional ambulation. Watch the child trying to crawl. Crawling eliminates the effect of deformities. Inability to crawl means that correcting the deformities would not help towards walking.

B	Factors to consider in surgical selection
	Selective motor control
	Balance
	Cognitive function visual impairment
	Apraxia
	Sensation
	Neurological involvement level



Good balance and presence of selective motor control are positive prognostic criteriae for orthopaedic surgery.



Results of orthopaedic surgery will not be satisfactory if the child does not understand what is expected of her or does not have the motivation to move.

E	Gross Motor Function Classification System (GMFCS)	
	Class	*Goal of treatment*
	1- Walks independently, speed, balance & coordination reduced	Diminish energy expenditure, decrease level of support, improve appearance
	2- Walks without assistive devices but limitations in community	
	3- Walks with assistive devices	Improve gait, position for sitting, transfers, supported standing
	4- Transported or uses powered mobility	Decrease pain, improve sitting & standing
	5- Severely limited, dependent on wheelchair	Better positioning, decrease pain, improve hygiene

A

Treatment according to stage of contracture	
Problem	*Treatment*
Dynamic spasticity, no contracture	Botulinum toxin, orthosis, stretching exercises
Muscle contracture but no joint deformity	Casting, muscle lengthening
Muscle contracture leading to joint deformity	Muscle lengthening, bone surgery

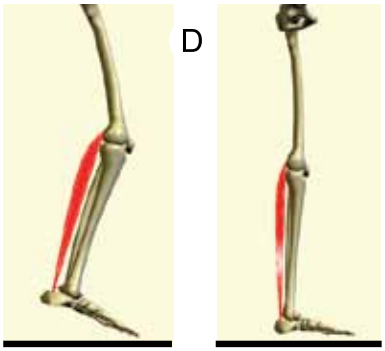
B

Types of CP	Surgical procedures most often performed
Quadriplegic	Hip adductor flexor release, osteotomy
	Spine fusion
Diplegic	Hamstring-gastrocnemius lengthening
	Hip adductor-flexor lengthening
	Derotation osteotomy
	Rectus femoris transfer
Hemiplegic	Gastrocnemius lengthening
	Split tibialis anterior & posterior transfer
	Tibialis posterior lengthening

C

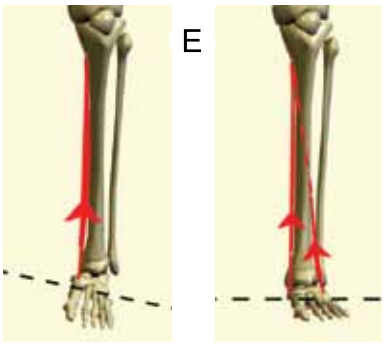
Aims of surgical procedures	
Tendon lengthening	Weakens spastic and shortened muscles, balances muscle forces
Split transfer	Balances deforming forces
Simple tenotomy	Balances deforming forces
Angular osteotomy	Corrects varus and valgus deformities of the foot & flexion deformities in the lower extremity
Hip surgery	Stabilizes the subluxated or dislocated hip
Rotational osteotomy	Corrects torsional deformities of the tibia or femur
Arthrodesis	Corrects deformity and stabilizes joints
Spine surgery	Corrects spinal deformity

D



Muscle lengthening: lengthen the short gastrocnemius muscle to achieve a plantigrade foot.

E



Split transfer of the posterior tibialis muscle balances the forces across the foot and corrects the varus deformity.

Orthopaedic interventions

When treating muscle contractures and deformities distinguish the stage of the problem and plan treatment accordingly [A].

Corrective casting

Corrective casting is used for minor ankle equinus contracture that does not respond to physical therapy or botulinum toxin injections; and knee flexion deformities that involve more than just hamstring tightness. A turnbuckle or hinged cast may help correct some significant knee flexion contractures. Apply the cast in a serial manner to the lower extremity with the knee and ankle as close to the anatomical position as possible. Local heat followed by vigorous stretching exercises are helpful beforehand to obtain better correction. Remove the cast and reapply if possible under sedation in 3 to 7 day intervals for 3 or 4 times.

The value of casting is controversial. Good results are possible over a long term. The effects may wear off after a few months. The compliance with serial casting is low due to the difficulties of repeated casting and cast removals. Some authors propose that casting weakens the already weak spastic muscles, creates atrophy and does not allow the antagonist muscle to work. However, it still has a place in the treatment of minor deformities of the knee and ankle. It is used after botulinum toxin injections as well.

Surgical methods

Orthopaedic surgical procedures used in CP are muscle releases and lengthenings, split tendon transfers, osteotomies and arthrodeses [B,C]. Upper extremity surgery is much more complex and should only be done by surgeons experienced in this field.

Muscle - tendon surgery Muscle-tendon lengthening [D] is the most commonly used method. It weakens spastic and shortened muscles, thereby balancing the forces acting on the joint. Split tibialis anterior and posterior tendon transfers of the foot help balance the deforming forces [E]. Simple tenotomies may be performed in selected muscles.

Osteotomy corrects varus and valgus deformities of the foot and flexion deformities in the lower extremity. Hip osteotomy stabilizes the subluxated or dislocated hip. Rotational osteotomies correct the torsional deformities in the tibia or the femur. Arthrodesis corrects deformity and stabilizes the joint.

Spinal fusion and instrumentation corrects spinal deformity.

Neurectomies are rarely performed.

Preoperative assessment

Evaluate all patients thoroughly before elective surgery to prevent complications or unpredictable outcomes. The motivation of the child and family is crucial to the success of the operation. Consider the family's resources (time, finance, access to therapy and hospital) for postoperative follow-up and

rehabilitation. Assess the severity of the problem to determine the expected functional result of surgery. The Gross Motor Classification System is a way to assess severity of involvement. Try to get treatment for co-morbidities such as seizures, gastro-esophageal reflux and infections preoperatively. Evaluate the severity of mental retardation, behavioural disturbances and social problems. Consider gastrostomies for children with oromotor dysfunction and growth retardation. Plan preoperative physiotherapy, exercises and instructions in walker or crutch use.

Postoperative care

Analgesia

Pain accompanies all operations. Focus on pain and anxiety relief, muscle relaxation in the immediate postoperative period [A]. Usually a combination of a narcotic analgesic and diazepam helps to control the immediate pain. Elevation of the extremity is essential for both pain relief and oedema prevention.

Pain is very common in the postoperative period, though it is difficult to predict who will have mobilisation problems because of pain. Postoperative pain management is important for early rehabilitation. Parenteral analgesics and patient controlled epidural analgesia are options to control pain [B]. Consider epidural anesthesia for all operations of the lower extremities in the older child. Insert a urinary catheter before the epidural. Continue the epidural catheter for postoperative analgesia at least 24 hours for muscle and 72 hours for bone operations. Turn the epidural off afterwards and switch to oral medications like diazepam or oxycodone. Remove the epidural catheter if oral medications control the pain. Routinely use antiinflammatory medication for pain relief. Do not use ketodolac after bony surgery because it delays bone healing. Reassure the child and family that the pain will subside within few days.

Ice application after all operations relieves pain and keeps the swelling down. Apply the ice for 2-3 days through the cast.

Try to distinguish spasticity pain from inflammatory muscle/joint pain and make an effort to relieve it. After surgery spasticity may be reduced because the muscle length has changed but it frequently returns 3 - 6 months later. Choose diazepam or baclofen for muscle relaxation and analgesia. Baclofen has analgesic effects in pain due to spasticity. The administration of diazepam as a central muscle relaxant and sedative agent is helpful.

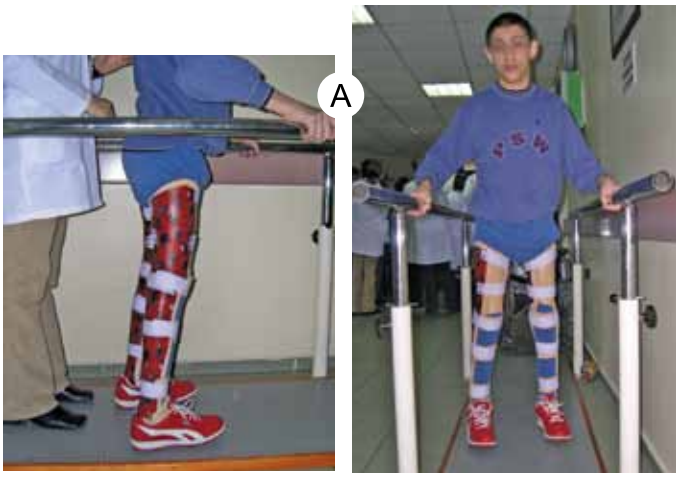
Mobilization

Minimize recumbency and immobilization. Encourage early mobilization and early weight bearing as well as strengthening of the trunk and upper extremities. Following surgery use casts [C], splints, plastic AFO's or KAFO's depending on the age and cooperation of the child as well as surgical stability. Choose prefabricated knee immobilizers in older children after hamstring lengthening. Allow to bear weight on the second to fourth days after soft tissue surgery. Time for weight bearing is guided by the quality of internal fixation of the bones in combined soft tissue and bony surgeries. Adequate nutrition and skin care are necessary to prevent complications such as pressure sores. Use a synthetic cast if the child has bladder control. Prefer a plaster of Paris cast and overwrap with synthetic material to avoid making the cast too heavy if the child is incontinent. A thin layer of plaster of Paris absorbs urine and prevents skin irritation.

Epidural catheter provides adequate analgesia and allows early mobilization.

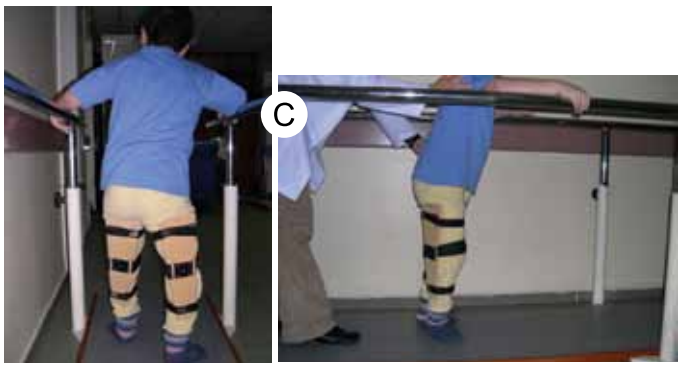
Postoperative medication (B)
Pain control
Epidural catheter
Antiinflammatory medication
(No ketodolac after bone surgery)
Reassurance & support
Ice over the cast
Spasticity control
Baclofen (oral)
Diazepam (oral or parenteral)

Immobilize the patient in a cast or brace for a few days after surgery. Osteotomies take longer to heal. Keep the child in cast longer after osteotomies. Elevate the legs with pillows under the shins. Use abductor wedges in the early postoperative period after hip adductor lengthenings.



Early mobilization is crucial for the success of surgery. Allow ambulation in plastic solid KAFOs in the 2nd day in patients who have had both hamstring and gastrocnemius lengthenings. Proceed to AFOs during the day after 3 weeks. Use night KAFOs for at least 6 weeks.

The knee immobilizer is lighter and easier to use than the plastic KAFO; however it is not suitable in severe flexion contractures.



Use either a prefabricated elastic knee immobilizer or a simple posterior shell after hamstring lengthenings without gastrocnemius surgery. The child will cooperate much better if his feet are free.

The hip Encourage prone sleeping early on after flexor releases or osteotomies of the hip. Have the child sleep in knee extension splints for 6-8 weeks after surgery to maintain hip extension.

Use an abduction wedge or pillow after adductor releases to keep the hips in 30º abduction at night. Continue postoperative abduction pillows for 6 weeks. Limit the time the child spends in a wheelchair after hip adductor and flexor releases or osteotomies. Do not allow sitting for more than an hour after bone surgery. Start active exercises on the 3rd postoperative day. Stretching exercises are essential.

Apply a hip spica cast for 4 weeks in young children after pelvic osteotomies, 3 weeks after femoral varus derotation osteotomies. There is no need for hip spica casts after intertrochanteric osteotomies with stable internal fixation above age 8. Bed rest is sufficient. Begin ambulation with crutches in the early postoperative period.

The knee Use cylindrical casts or splints in knee extension for 3 weeks after hamstring lengthening. Have the child wear the splints [A] or knee immobilizers [B] at night for 6 weeks after surgery. Limit elevation of the leg in bed to one pillow in order to prevent stretch of the sciatic nerve. Mobilize the patient within 2 or 4 days with the cast or splint on.

Immobilize the knee in an extension brace or knee immobilizer after rectus femoris transfers. Begin knee flexion exercises and weight bearing on the second to fourth postoperative days to prevent adhesions and knee stiffness [C].

The foot The most common surgery in the foot is heel cord lengthening. In a very young child use a Vulpius type procedure. An AFO is usually sufficient after surgery. For tendon surgery i.e. split tendon, total tendon or anterior tibial tendon transfers, recommend 6 weeks in a walking cast. For bone operations such as calcaneal lengthenings, subtalar arthrodesis and osteotomies of the cuneiform bones, recommend 6 weeks in a cast; non-weight bearing for the first 3 weeks, weight bearing for the second 3 weeks.

The spine Following spine fusion for scoliosis spinal braces are not necessary if the fixation is stable. Use body casts or braces for 6 months in children with poor bone quality.

Night splints A period of splint use to prevent the recurrence of contractures after cast removal is helpful. However, splints disturb sleep by constantly keeping the muscles in the stretched position. Night splints should be used very carefully because not all children will tolerate them. It is important that the child sleeps well. Splints are not necessary in children whose muscle tone decreases during sleep. The correction obtained by splints should not be painful for the child. A way to improve compliance is to use night splints on one extremity the first night and on the other the next night.

Children who tend to have recurrence of equinus or who have had a second heel cord lengthening should sleep in a dorsiflexion splint at night. The severe quadriplegic child who has dystonia and tends to contract severely can be placed in an A frame at night. Do not use night time knee extension splints for more than 3 months post operatively.

Postoperative physiotherapy

A significant change in all the primary impairments is expected after surgery. There is a need for gentle return to function. Try to regain range of motion and strength as early as possible after surgery. Begin mobilisation as soon as the child is comfortable and painless, usually on the second to fourth day after soft tissue procedures. Do not allow weight bearing for 3 weeks after osteotomies.

Begin training with range of motion exercises and gradually progress to strengthening as healing allows. Keep in mind that a spastic muscle is also a weak muscle. Strengthen the muscles after muscle lengthening. The ultimate aim is to improve the ambulatory capacity. It usually takes approximately 3 months to regain the preoperative muscle strength after multilevel surgery. Immediate postoperative physiotherapy re-introduces movement and the new alignment. The skills that the patient acquires are established in 3-6 months after surgery. Provide intensive physiotherapy in this period. Monitor changes in the patient's status attributable to growth or increased spasticity 6-12 months postoperatively. Change in function will not be very obvious for up to one year after the operation.

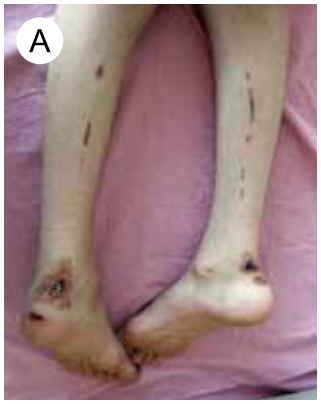
Physiotherapy ends when the child has no more change in strength, function and skill level. Other ways to provide therapeutic movement programs for these children such as sports and play are encouraged. The rehabilitation physician must also monitor for new dynamic or fixed contractures, orthotic problems, prescribe accurate braces and provide guidance for adaptive equipment.

The 'Birthday Syndrome'

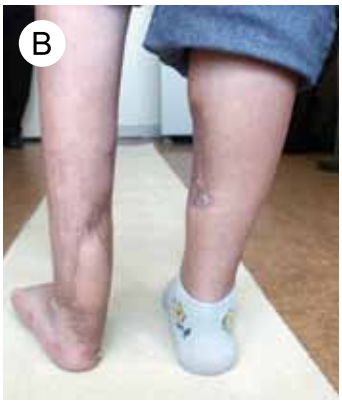
One group of complications related to multiple operations over the years is social isolation, loss of motivation, frustration and psychosocial problems. Do not perform a chain of surgical operations leading to 'The Birthday Syndrome' [A] as described by M. Rang. Plan to do all necessary surgical interventions at the same time if the child's medical and social status permits. This is called single event multilevel surgery and spares the child the burden of multiple consecutive surgical interventions throughout his life. Do not forget, however, that treatment plans must be individualized for each child according to his specific needs. Tailor treatment for each patient.

Illustration by Idil Çilingiroğlu

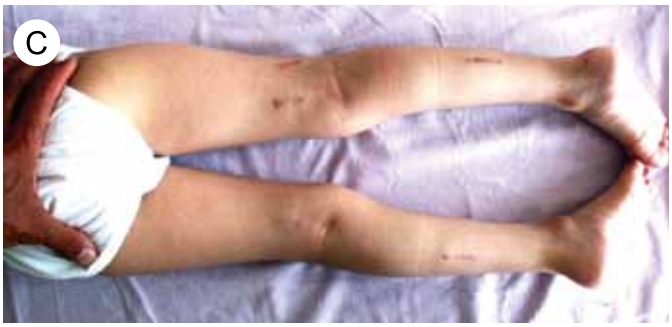
The 'Birthday Syndrome': The child who has an operation every year has to spend all his birthdays recuperating from surgery in casts.



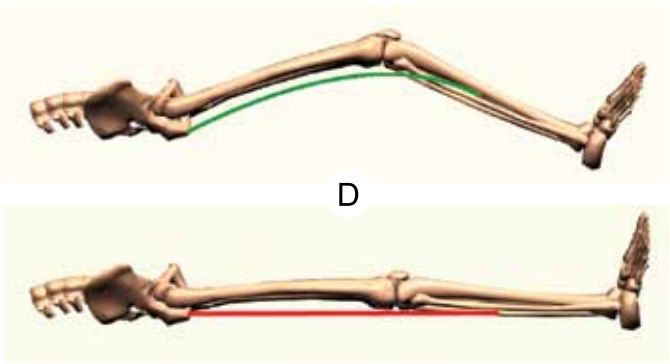
Pressure sores can occur when the child is in the cast. Cast saws can also create psychological and physical trauma. Bivalve the cast in the operating room if possible to avoid further injury.



Tendo Achilles lengthening resulted in wound complications and skin necrosis in this elder hemiplegic child. Skin grafting and a cross leg flap was necessary.



Efficient technique leaves minimal scarring on the extremities even when multiple incisions are performed. The incision marks of six muscle lengthenings in this child are barely visible few months after surgery.



Sciatic nerve traction injury can occur after surgery to relieve knee flexion contracture. Avoid excessive elevation of the operated extremity postoperatively to minimise traction on the nerve.

Complications of surgery

Immediate postoperative complications such as infection, thrombosis and pulmonary embolism are very rare in young children. Complications of general anesthesia such as pneumonia or pressure sores from lying in the bed are possible. Pressure sores are common particularly in the malnourished children [A]. Children who have hip osteotomies and are treated without a spica cast in bed often will lie in one position and develop pressure areas on their heels, buttocks or the sacral region. It is important to make sure that there is a little elevation under the ankle so that the heel does not get pressure.

Achilloplasty incisions frequently cause skin lesions in elder patients [B]. For an uneventful recovery from orthopaedic surgery keep the incisions small and use intracutaneous resorbable sutures to minimize the trauma of suture removal later [C].

Always keep the risk of a postoperative fracture in mind in the immobilized and osteoporotic patients, particularly children who are wheelchair bound spastic quadriplegics. After a child has been immobilized in a spica cast the incidence of a supracondylar fracture has been reported to be as high as 20%. Be careful when mobilising the child after spica cast removal. Signs of fracture are swelling and pain in the distal femur. Obtain radiographs at the least suggestion.

Recently bisphosphonates have been used to treat osteoporosis in CP patients. These should be reserved for the most severely involved patients. They can be given either IV or orally. Many children who have spastic quadriplegia also have significant esophageal reflux problem and they will not tolerate the oral medication. Oral bisphosphonates can be harmful because it increases gastroesophageal reflux, ulcers and have other gastrointestinal side effects.

One of the significant complications that can occur is overlengthening of tendons. Heel cord overlengthening will lead to pes calcaneus which is worse than mild equinus. Hamstrings that are overlengthened can lead to recurvatum of the knee which is worse than mild flexion. Vigorous stretching of the knee in the supine position with the hip flexed to 90^{o} after hamstring lengthening can cause a neuropraxia of the sciatic nerve [D]. Therefore optimum range of motion following hamstring lengthening should not exceed 70^{o}.

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Anesthesia and Chronic Pain Management

Patients with CP frequently present for surgery for a variety of reasons. Common surgical conditions include lower and upper limb orthopaedic surgery, spinal surgery, dental procedures, surgery to control gastroesophageal reflux (reflux), gastrostomies and surgical procedures to control spasticity such as selective dorsal rhizotomy and intrathecal baclofen pump implantation [A]. The patient with CP presents a real challenge for anesthesiologists because of the associated multiple disabilities and systemic problems. A pediatric anesthesiologist who has experience with children with neuromuscular disorders is an asset to the treatment team.

Preoperative assessment

Children with CP require special consideration because of their various disabilities [B]. Medical, communication, general care problems and social issues often complicate the preoperative assessment of patients with CP.

Be aware of the visual and hearing deficits, behavioural and communication problems that the child has.

Behavioural problems

Children are sensitive and need enough time to communicate their feelings and problems. The parental attitudes must be accepted and respected. Make every effort to increase communication with the child and the parents. Be understanding of the aggressive, overprotective or negative behaviours they show, recognize that these are manifestations of stress, fear, anger and frustration.

Gastroesophageal problems

Evaluate all systems extensively to ensure an uneventful perioperative course. Esophageal dysmotility, abnormal lower esophageal sphincter function, and spinal deformity can cause reflux and pulmonary aspiration. Cisapride, commonly used for treatment of reflux, can cause prolonged QT interval and ventricular dysrhythmias. Drooling can be a problem during induction and emergence from anesthesia.

Nutritional problems

Electrolyte imbalance and anemia from malnutrition require preoperative nutritional support in a small group of patients with feeding problems. Consider preoperative feeding gastrostomy to improve nutrition, decrease postoperative infection risk, and enhance wound healing.

Pulmonary problems

Start preoperative physiotherapy, bronchodilators or antibiotics as required to optimize the perioperative care of patients with recurrent respiratory infections and possibility of pulmonary aspiration from reflux and chronic lung disease.

Dental and mouth hygiene

Note the presence of loose teeth, dental caries and temporomandibular joint dysfunction preoperatively because these frequently cause difficulty with laryngoscopy during endotracheal intubation.

Epilepsy

Children who receive regular anticonvulsant medication should continue their medicine during the perioperative period for symptom stability. Keep in mind the side effects of these agents, such as bleeding tendency with valproate sodium.

Latex allergy

Patients with CP are at increased risk of developing latex allergy because of multiple surgical procedures and exposure to latex allergens from an early age. Ask the parents about respiratory symptoms such as wheezing or allergic rhinitis and cutaneous manifestations such as rush, itch, oedema when exposed to products containing latex.

Spasticity

Remember that antispastic medications such as baclofen, benzodiazepines and botulinum toxin have side effects that interfere with anesthesia. Oral baclofen may delay emergence from anesthesia and cause bradycardia-hypotension during anesthesia.

Premedication and preoperative management

Forbid the intake of solid foods 4 hours before surgery in infants and 8 hours before surgery in older children. Allow clear fluids and water until 3 hours before surgery.

The child with CP may be anxious because of the past multiple hospital admissions. He is frightened and does not understand what is expected. Show empathy and patience during induction. Consider sedatives to reduce anxiety and spasm. The response to such medication may be unpredictable. Psychological delay and behavioural disturbance result in serious separation anxiety. Allow the parents to remain with the child during the induction and recovery periods to minimize this anxiety [C].

Increased tone may prevent the insertion of an intravenous line in the spastic or dystonic child considering that stress and fear increases muscle tone even more. Apply creams containing lidocaine or prilocaine to the venepuncture site at least 1 hour before insertion to lessen trauma and ease induction.

Common surgical procedures
Lower and upper limb orthopaedic surgery
Spine surgery
Dental procedures
Anti-reflux surgery
Gastrostomy
Selective dorsal rhizotomy
Intrathecal baclofen pump implantation

A

Special considerations in preoperative assessment
Behavioural problems
Gastroesophageal problems
Nutrition
Pulmonary
Dental and mouth hygiene
Convulsions
Latex allergy
Spasticity

B

C

Allow the mother into the operating theatre to minimize the child's anxiety.

Cover the young child in cotton to minimise heat loss during surgery.

Depending on the extent of surgery caudal or epidural analgesia will provide effective pain relief.

Positioning for surgery may be difficult due to contractures, bilateral tourniquets and multiple catheters.

Contractures prevent appropriate positioning and cause increased pressure over the bony prominences. Skin ulcers occur especially if epidural analgesia is used.

Anesthetic management

Anesthesia is difficult because of lack of communication and cooperation with the patient. Intravenous access may be a problem caused by spasticity, dystonia or refusal. Consider rapid sequence induction especially in patients with reflux. Inhalational induction of anesthesia may be messy and risky in patients severely affected by drooling. All common agents for intravenous induction of anesthesia are suitable but propofol seems to be more painful for these patients.

Secure the airway with a laryngeal mask when clinically appropriate. Perform tracheal intubation if reflux and drooling are present. A nasogastric suction catheter may be beneficial for longer operations.

Studies of muscle relaxants indicate slight sensitivity to succinylcholine and resistance to vecuronium in patients with CP. The possible mechanisms include drug interaction with anticonvulsant medication and chronic immobilization.

Minimum alveolar concentration for anesthesia may be decreased for volatile anesthetics. Most anesthetics have anticonvulsant properties. Avoid enflurane, ketamine and etomidate in patients with epilepsy.

Perioperative heat loss is a common significant problem causing systemic problems and delaying emergence. Minimize hypothermia by humidifying gases, using warmed intravenous fluids, using forced air warming blankets and avoiding a cold operating room [A].

Perioperative epidural anesthesia decreases the requirement of general anesthetics and the incidence of delayed arousal from anesthesia [B].

Proper patient positioning may be difficult because of contractures [C]. Consider taking precautions for prevention of pressure sores and nerve damage [D].

Insert a urinary catheter for long operations with significant blood loss and fluid shifts to monitor urine output and decrease the risk of renal failure that is caused by inappropriate fluid administration.

Postoperative management

Emergence from anesthesia may be delayed because of hypothermia and residual volatile anesthetic agents. Frequent suctioning is required in patients who had been drooling preoperatively. Protect the airway from excessive secretions, regurgitation and vomiting.

Patients will be irritable [E] during emergence from anesthesia in unfamiliar surroundings, especially if they have intellectual disability, pain, visual defects, hearing deficits or urinary retention.

Mother's care and affection help a lot during early recovery.

Continue intrathecal baclofen during the perioperative period to prevent abrupt withdrawal symptoms such as convulsions, hallucinations and dyskinesias. Maintain anticonvulsant medication for children with epilepsy. During the early postoperative period diazepam can be administered rectally, sodium valproate may be given IV in countries where this preparation is available.

Decreased mobility, inefficient fluid intake and loss of peristaltism increase the tendency to develop constipation. Oral, intravenous or epidural opioids contribute to this problem.

Pain and spasticity

Focus on relieving pain, anxiety and muscle spasm. Postoperative pain management is crucial for the success of surgery. Epidural analgesia diminishes both postoperative pain and muscle spasm. It is accepted as the gold standard for pain management in patients with CP. Epidural administration of opioid and local anesthetic combination (bupivacaine+fentanyl) is the most effective and safest method for postoperative analgesia because it causes less sedation. Add clonidine to achieve better control of muscle spasm. Epidural analgesia has certain advantages and dysadvantages [A,B]. A common side effect of epidural analgesia is urinary retention. Insert a Foley catheter preoperatively to prevent postoperative urination problems. Leave the epidural catheter in place for 2 to 3 days after soft tissue procedures and 4 to 5 days in cases of bone surgery. Turn off the infusion and start oral medications such as diazepam or oxycodone after 5 days. Remove the epidural catheter if oral medications control the pain. For the young child who has minor soft tissue surgery or simple hip plate removal, it is not necessary to use an epidural catheter. A postoperative caudal block will allow such patients to emerge from anesthesia without pain.

Nonsteroidal antiinflammatory analgesics or paracetamol is the second choice after epidural analgesia. Rectal paracetamol (40 mg/kg loading dose, then 20 mg/kg/ four times daily) will relieve postoperative pain in most cases. If not sufficient, consider parenteral opioids (morphine, tramodol and meperidine).

Postoperative muscle spasms are a common and serious problem. Epidural analgesia is advantageous in children undergoing lower extremity surgery because it decreases reflex muscle spasms triggered by pain. For patients in whom epidural analgesia is not appropriate, benzodiazepin (diazepam 0.1- 0.2 mg/kg or midazolam 0.05 - 0.07 mg/kg) administered orally, rectally, or intravenously can be used to alleviate spasm but often results in prolonged sedation.

An alternative drug for the management of postoperative spasticity is oral baclofen. Baclofen has analgesic effects on pain caused by spasticity.

Chronic pain management

Chronic pain results from hip instability, gastro-esophageal reflux, menstrual discomfort in adolescent girls, back pain caused by scoliosis, facet joint arthritis, capsulitis and spasticity. It is difficult to determine the cause of and treat chronic pain in the child who has a communication disorder [C]. A multidisciplinary approach is necessary with the team members ready for consultation, evaluation and treatment on the same day if possible. Depression during the adolescent period can increase chronic pain. Patients with CP are undertreated for pain because of various reasons [D].

A

Advantages of epidural analgesia
The most effective analgesia
Decreased muscle spasm
Early mobilization
Decreased pulmonary complications
Decreased risk of deep venous thrombosis (not a major issue in the young child)

B

Dysadvantages of epidural analgesia
Urinary retention
Side effects of opioids (Itching, nausea, vomiting)
Increased risk of pressure ulcers or compartment syndrome
Complications Spinal hematoma Nerve root damage Infection Dural puncture

Differential diagnosis of chronic pain and restlessness in the quadriplegic patient

Etiology	*Symptom*	*Treatment*
Hip	Pain during transfers Night pain Less with position change	Analgesics (paracetamol), NSAID, intraarticular steroids Surgery
Reflux	Anemia	Antacids, ranitidine
	Weight loss	Proton pump inhibitors
	Anorexia	Erect posture, proper sitting position
Low back pain	Pain after sitting for a longtime	Physiotherapy
	Night pain	Tricyclic antidepressants
		Paracetamol, NSAID,
		Facet joint injections
		Wheelchair modifications
Menses pain	During menses	Oral contraceptives

C

D

Causes of undertreatment for pain
Too many medical problems that may cause pain
Frequent painful procedures
Behavioural problems that mask expression of pain
Difficult to interpret pain indicators because of physical problems
Comfort of the patients valued less by society

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Pathophysiology of Spasticity

Spasticity is a major neuromuscular problem in CP. It is so deeply engrained in medical and public literature that a spastic child has come to mean a child with CP for most people around the world. Spasticity is difficult to define. The pathophysiology is obscure, findings on examination are inconsistent, and treatment is not always successful. Understanding the physiology of normal movement may help the physician in the management of spasticity.

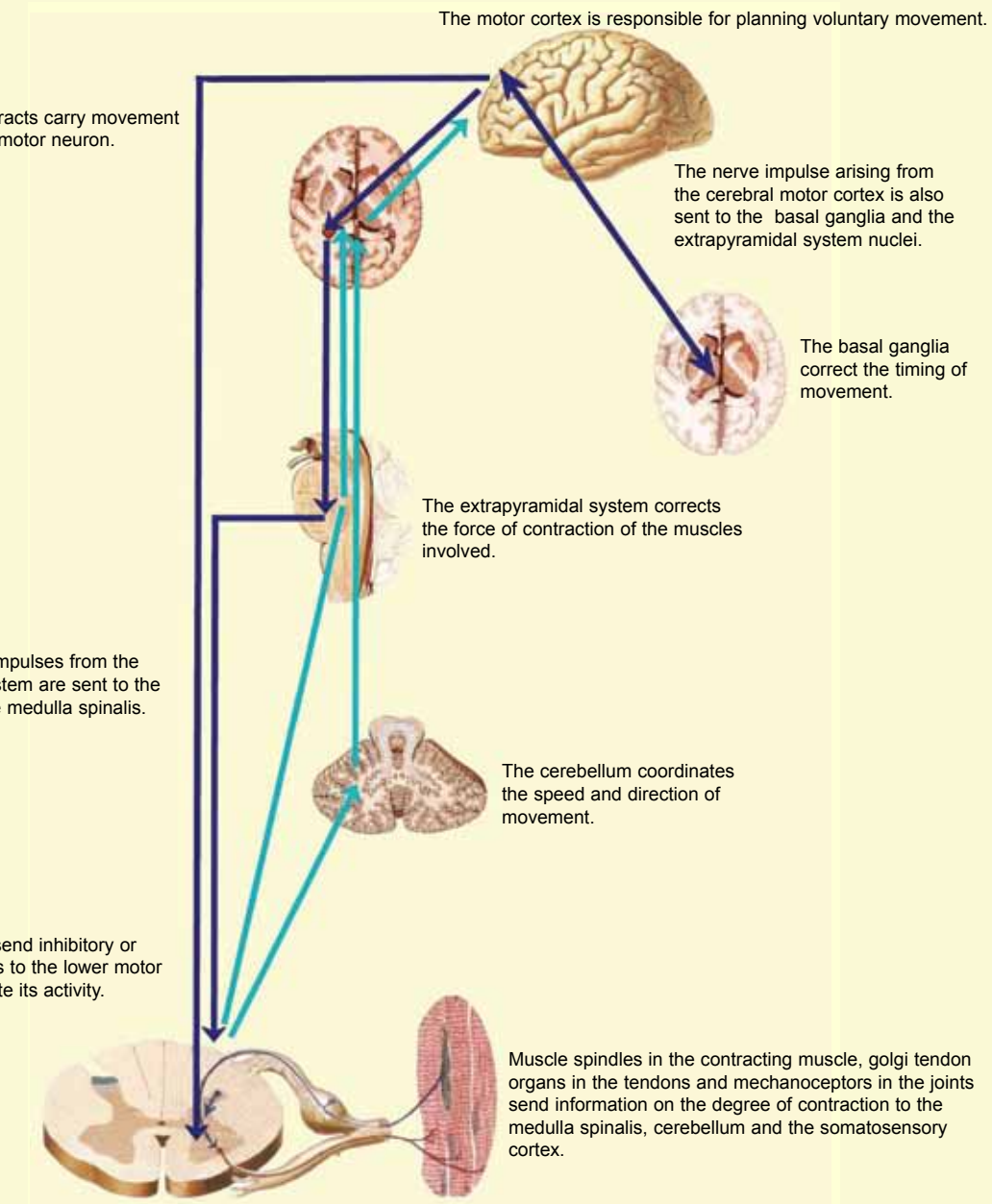
Physiology of movement

Afferent input from the internal organs, the musculoskeletal system, and the skin converge on the medulla spinalis. This afferent input activates the stretch reflex, both directly and through the interneuron, and results in a reflex motor response [A].

The same afferent information goes to the cerebellum and the somatosensory cortex. It is processed in those centers as well as in the basal ganglia. The resulting motor response is relayed to the lower motor neuron through the pyramidal and extrapyramidal system tracts. The pyramidal tracts go directly to the lower motor neuron whereas the extrapyramidal tracts

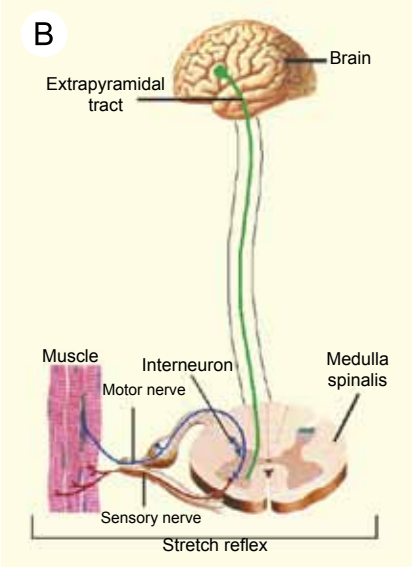
A

Neural pathways regulating muscle contraction



The upper motor neuron syndrome (A)
Positive findings
1. Increased muscle tone
2. Exaggerated tendon reflexes
3. Clonus
4. Babinski positive
5. Flexor synergies
Negative findings
1. Loss of selective motor control
2. Loss of hand and finger dexterity
3. Muscle weakness
Results in muscle
1. Stiffness
2. Contracture
3. Fibrosis
4. Atrophy

Table modified from: Mayer NH: Clinicophysiologic concepts of spasticity, Spasticity: Etiology, Evaluation, Management and the Role of Botulinum Toxin. Eds. Mayer NH, Simpson DM, WEMOVE, 2002



The hyperexcitability of the lower motor neuron is presumed to be the cause of spasticity. This hyperexcitability is evident in the increase in deep tendon reflexes.

Modified Ashworth Scale (C)	
0	No increase in muscle tone
1	Slight increase in muscle tone, manifested by a catch and release or by minimal resistance at the end range of motion when the part is moved in flexion or extension/abduction or adduction, etc.
1+	Slight increase in muscle tone, manifested by a catch, followed by minimal resistance throughout the remainder (less than half) of the ROM
2	More marked increase in muscle tone through most of the ROM, but the affected part is easily moved
3	Considerable increase in muscle tone, passive movement is difficult
4	Affected part is rigid in flexion or extension (abduction ,adduction, etc.)

end at the interneuron. The cerebellum, basal ganglia, and extrapyramidal system nuclei modify the motor response as it goes to the medulla spinalis. In this way all motor output is influenced by the incoming sensory input and converges on the lower motor neuron. The interneurons in the medulla spinalis regulate the activity of the motor neuron.

The upper motor neuron syndrome

CP results in an upper motor neuron syndrome [A] characterized by spasticity, exaggerated tendon reflexes, clonus, pathological reflexes, mass synergy patterns, muscle weakness, loss of selective motor control and loss of hand dexterity. Spasticity is a component of the upper motor neuron syndrome.

Definition of spasticity

Muscles show a physiological resistance to passive motion. This is called muscle tone. Spasticity is the increase in this physiological muscle tone. The terms "spasticity" and "increased tone" may be used interchangeably. Spasticity is velocity dependent. The faster the passive movement, the greater the resistance of the muscle. The increase in muscle tone causes loss of trunk balance and difficulty of active movement in the extremities.

Pathogenesis

The pathogenesis of spasticity is presumed to be an increase in the excitability of the lower motor neuron. This presents as hyperactive stretch reflexes [B] on clinical examination. Many hypotheses attempt to explain this hyperexcitability. One suggests a change in the balance of excitatory and inhibitory inputs to the motor neuron pool. When the inhibitory inputs are reduced, the interneurons send excitatory impulses to the lower motor neurons and they become hyperexcitable.

Measuring spasticity

Spasticity can be measured by clinical examination, mechanical instruments, and electrophysiological techniques [C]. The modified Ashworth and Tardieu scales are commonly used for clinical evaluation. They measure tone intensity but do not evaluate the effect of spasticity on function. Mechanical instruments measuring the resistance of the muscle to passive stretch and electrophysiological measures showing the hyperexcitability of the stretch reflex are used only for research purposes.

Measurements in spasticity (D)
Clinical measures
Range of motion
Tone intensity measures
Modified Ashworth Scale
Tardieu Scale
Mechanical instruments
The pendulum test
Electrophysiological measures
The H reflex
Vibration inhibition index
Functional measures
Upper extremity function
Gait

The Ashworth scale The Ashworth scale [D on previous page] is by far the most commonly used evaluation method for spasticity. Always test the patient while he or she is in a relaxed supine position. Passively move the joint rapidly and repeatedly through the available range of motion and grade the resistance using the definitions.

The Tardieu scale The Tardieu scale measures the intensity of muscle tone at specified velocities [A]. Always grade the Tardieu Scale on the same day. Keep the body in a constant position for a given extremity. Keep the other joints, particularly the neck in a constant position throughout the test and from one test to another. Perform the test at a reproducible velocity of stretch. Note the joint angle at which the catch is first felt.

Determine the effect of spasticity on the child's function, ease of care and quality of life by using various functional scales. Effect of spasticity on function and well being should guide treatment.

Tardieu scale A

Quality of muscle reaction is measured as:

0	No resistance throughout the course of the passive movement
1	Slight resistance throughout the course of the passive movement
2	Clear catch at precise angle, interrupting the passive movement, followed by release
3	Unsustained clonus (less than 10 sec when maintaining the pressure) occurring at a precise angle, followed by release
4	Sustained clonus (more than 10 sec when maintaining the pressure) occurring at a precise angle

Angle of muscle action is measured relative to the position of minimal stretch of the muscle (corresponding to angle zero) for all joints except the hip where it is relative to the resting anatomical position.

Effects of spasticity

Adverse effects Spasticity causes [B] difficulty in movement, abnormal posture in sitting and standing, contractures leading to deformities, pressure sores and pain. Increase in tone is uncomfortable. Sitting is difficult for the nonambulatory child because of increased adductor and hamstring muscle tone. The child slides out of the wheelchair and cannot be positioned properly. He cannot transfer to and from the bed, wheelchair and bathtub. Perineal hygiene and dressing the child require more effort. The ambulatory child has trouble initiating movement. He cannot wear his braces. Energy cost of movement increases. Patient loses function and parents have difficulty caring for the child.

When muscle tone increases, muscles become tight. This inhibits normal gait and posture. Normal movement patterns do not develop. Instead, the child shows abnormal or compensatory movement patterns. Spasticity affects muscle growth. Muscles need to be stretched while relaxed; failure to do this results in poor growth. Spasticity initially causes apparent muscle shortening but the passive range of motion is full. This abnormal permanent resistance is dynamic contracture. If uncorrected, fibrosis and eventually bony deformity lock the joint into a fixed contracture. How fast a contracture will develop depends on the severity of spasticity and the muscles involved: contractures progress more quickly in some muscles.

Bone growth is distorted by the abnormal resistance of the shortened muscles. Growing bone easily gives way to sustained pressure. Untreated spasticity causes excessive stress on bone that produces abnormal rotation. This stress may also inhibit physiological derotation of long bones. If spasticity is not relieved at an early stage, bone deformities occur. Prolonged equinovarus caused by triceps surae and tibialis posterior spasticity might rotate the tibia inwards. Spasticity of hip adductors can rotate the femur inwards, thus inhibiting the physiological derotation process of infantile femoral anteversion.

Beneficial effects Increased tone may be useful for the child. It helps maintain to keep the legs straight, thereby supporting the child's weight against gravity. The child with increased tone in trunk extensors may stand and take a few steps. Spasticity may help preserve muscle bulk and bone density.

Effects of spasticity B

Positive effects	
	Extensor tone in the limbs help standing
	Preserve muscle bulk
	Preserve bone density
Negative effects	
	Masks contraction in the antagonist
	Difficulty in movement
	Abnormal posture
	Difficulty in sitting and transfers
	Inhibits muscle growth
	Leads to contractures
	Difficulty in hygiene and dressing
	Pressure sores
	Pain

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Goals of spasticity treatment
Increase function
to perform better in activities of daily living
to walk better
Increase sitting ability and balance
Prevent deformity & decrease contractures
Pain relief
Improve hygiene and patient care

A

Treatment methods
Physiotherapy
Positioning
Exercises
Stretching
Neurofacilitation
Electrostimulation
Splinting & Casting
Oral medications
Baclofen
Diazepam
Clonazepam
Dantrolene
Tizanidine
Intrathecal medications
Baclofen
Morphine
Clonidine
Neuromuscular blocks
Local anesthetics
Phenol
Botulinum toxin
Orthopedic surgery
Selective dorsal rhizotomy

B

Have the child sit with legs in front, knees extended and ankles in neutral to stretch the hamstring and gastrocnemius muscles. This position is difficult to maintain for long periods.

Essentials of Spasticity Treatment

Indications for treatment

Consider treating spasticity when it causes loss of function or produces contractures, deformities, pressure sores, or pain [A]. Additional indications include difficulty in positioning or caring for the total body involved child. Even though a wide range of treatments exist, none of them is fully satisfactory. Unwanted side effects limit the use of certain modalities. Some children do not respond to any of the antispasticity measures. The success of treatment depends on having specific goals in treatment, choosing the correct method according to the child's problem and monitoring for side effects and complications.

Treatment methods

Treatment options are divided into reversible and permanent (surgical) procedures [B]. They can also be classified as systemic or local treatments. All treatment procedures aim to modulate the stretch reflex. In mild spasticity, basic measures such as positioning, exercises and bracing may be sufficient whereas in more severe cases, interventions can be more invasive. Often, treatments are combined to decrease side effects and to improve outcome.

Physiotherapy

Physiotherapy is a fundamental part of spasticity management. Muscle overactivity produces muscle shortening and muscle shortening increases spindle sensitivity. Muscle contracture and stretch sensitive muscle overactivity are intertwined. Therefore physical treatments aimed at lengthening the overactive muscles are fundamental. Address both shortening and overactivity. Consider applying various techniques such as positioning, ice, and exercises for these purposes.

Positioning Position the child to stretch the spastic muscles and decrease the sensitivity of the stretch reflex and the brain stem reflexes that trigger spasticity [C]. The therapists should teach these positions to the family so that the child lies and sits this way most of the time at home. Head supports may improve tone in the trunk muscles by providing a sense of safety and inhibiting the tonic neck reflexes. Advise use of the tailor-sitting position to reduce adductor spasticity [D]. Good seating provides a stable platform and facilitates good upper extremity function.

Stretching exercises Stretching muscles may prevent contractures and promote muscle growth. Spasticity decreases with slow and continuous stretching. This effect lasts from 30 minutes to 2 hours. Use stretching exercises before bracing and serial casting to obtain the necessary joint position.

D

Sitting in a cross legged position applies slow static stretch to the adductors and decreases spasticity.

Neurofacilitation techniques Most neurofacilitation techniques are used to reduce muscle tone [A]. With the Bobath method, the therapist positions the child in reflex inhibitor positions and provides kinesthetic stimulation to inhibit the primitive reflexes and elicit advanced postural reactions to normalize muscle tone. With the Vojta method [B], different positions and proprioceptive stimulation are used for the same effect. Tone reduction lasts for a relatively short period of time with both methods.

Inhibitive (Tone Reducing) Casting and Bracing

Muscle relaxation after stretching exercises lasts for a short period of time. For longer duration the stretch on the muscle should be maintained for several hours every day. This is possible with the use of rigid splints or serial casting [C]. The effects are maximal if the cast or the splint is applied after the muscle is relaxed.

The tone-reducing effect of casts and splints is controversial. Some think that casts decrease muscle tone by creating atrophy in the already weak spastic muscle. Casts also cause pressure sores in children who are malnourished and have severe spasticity. Patient compliance may be poor because of difficulties of living with the cast.

Consider casting as an adjunct to treatment with local antispastic medications in the young diplegic or hemiplegic child with severe spasticity interfering with ambulation to delay orthopaedic surgery.

At present, the most common methods of spasticity management in cases of CP are oral medications, botulinum toxin, phenol or orthopaedic surgery [D].

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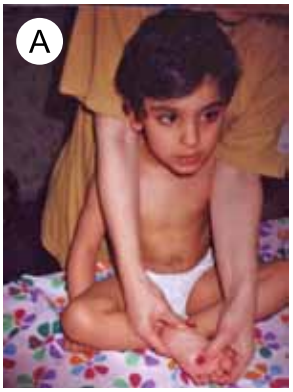
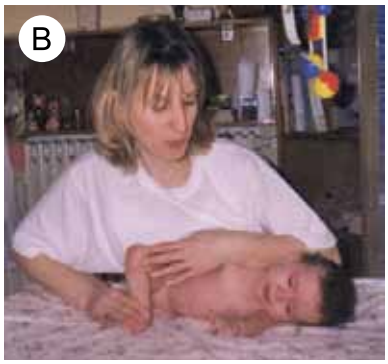


The Bobath method uses positioning and kinesthetic stimulation to reduce muscle tone.



The Vojta method has two basic positions of reflex rolling and crawling used both to initiate movement and to diminish muscle tone.

Long leg casts keep the knees in extension and the ankles at 90° flexion.

D

Treatment options in spasticity

	Age	*Patient group*	*Indication*	*Follow-up care*	*Result*	*Side-effect*
Oral medications	Any age 2-5 most common	Total body involved	Severe spasticity	Rehabilitation	Mild reduction	Sedation, weakness
Botulinum toxin A	Any age 2-10 most common	All spastic types	Focal spasticity too young for other interventions	Range of motion, stretching, strengthening exercises	Effective for 3-6 months good results in walking and ADLs	None obvious
Intrathecal baclofen	Above age 3 Abdomen large enough for pump insertion	Total body involved spastic or dystonic	Severe spasticity interfering with function or patient care	Range of motion exercises	Less need for orthopaedic surgery easier care better sitting	Infection Cerebrovascular fluid leak
Orthopaedic surgery	5-15 years	All spastic types	Contractures & deformities	Strengthening	Better walking	Recurrence, weakness
Selective dorsal rhizotomy	3-7 years	Diplegic patient with pure spasticity	Spasticity interfering with walking	Intensive physiotherapy	Controversial	Increasing scoliosis, hip instability risk of incontinence

Oral Medications

Various pharmacological agents decrease spasticity. Baclofen, benzodiazepines (diazepam, clonazepam), dantrolene sodium and tizanidine are commonly used in children [A].

Indications

Consider systemic oral antispastic drugs in total body involved nonambulatory children with generalized spasticity. They are also useful for short periods after orthopaedic surgery. Systemic side effects such as drowsiness, sedation, and generalised weakness are common, so they generally are not recommended for ambulatory children. Keep the initial dose low and gradually titrate to a level at which the effect is maximal and the side effects are minimal. The responses of children to oral antispastic drugs are not consistent. Try different drugs to achieve a satisfactory clinical effect.

Oral antispastic drugs

Baclofen

Baclofen is an agonist of the main inhibitory CNS neurotransmitter gamma aminobutyric acid (GABA). It shows its effect mainly on the spinal cord. It decreases spasticity by increasing the inhibitory effect of the interneuron on the alpha motor neuron. The lipid solubility of baclofen is poor, so it cannot easily cross the blood brain barrier. High oral doses are necessary to achieve a therapeutic dose in the cerebrospinal fluid (CSF). The effect starts 1 hour after ingestion and lasts for 8 hours. The drug must be taken three to four times daily in divided doses. Daily dose for children between ages 2 to 7 is 10 to 15 mgrs per day with a maximum of 40 mgrs per day. After the age of 8 years, the dose may be increased to 60 mgrs per day. Maximum doses range between 80 to 120 mg. per day in adults. Side effects including sleepiness, sedation, drowsiness, fatigue, headache, nausea, and a decrease in seizure threshold are commonly associated with increasing doses. Baclofen also causes generalised muscle weakness. All side effects are dose dependent. Sudden withdrawal may cause hallucinations and seizures sometimes accompanied by extreme hyperthermia and increased spasticity called the baclofen withdrawal syndrome. The dose of the drug must be decreased gradually.

Diazepam

Diazepam is a benzodiazepine tranquilizer that works as a GABA agonist. It enhances the presynaptic inhibitory effect of GABA and decreases spasticity. It is absorbed faster than baclofen, acts faster, and has a longer lasting effect. Doses in children range between 0.12 to 0.8 mg/kg body weight with a maximum of 20 mg. daily divided into two or three equal doses. Diazepam decreases painful muscular spasms and improves sleep. Sedation and other CNS side effects are very common, so this drug is not recommended for treating ambulatory children except after orthopaedic surgery when it improves the child's tolerance and participation in the rehabilitation program. CNS side effects are weakness, memory loss, ataxia, depression, and dependency.

Clonazepam

Clonazepam has an effect similar to that of diazepam, but it has a slightly longer half-life. It is preferred over diazepam because its side effects are fewer. Initial dose is 0.1 to 0.2 mg/kg/day. This dose is titrated for an optimal effect.

Dantrolene sodium

Dantrolene sodium inhibits muscle contraction by blocking calcium release from the sarcoplasmic reticulum in the muscle fiber. Initial dose is 0.5 mg/kg of body weight with a maximum dose of 3 mg/kg of body weight. Total daily dose should not exceed 12 mg per day administered in four divided doses. Side effects include muscle weakness, sedation, diarrhoea, and hepatotoxicity. CNS side effects are rare. Liver function tests should be performed two to four times a year, and the total treatment duration should not exceed 2 years.

Tizanidine

Tizanidine is an alpha adrenergic receptor agonist. It shows its effect at the brain and the spinal cord level. Tizanidine decreases the release of excitatory neurotransmitters and increases the release of inhibitory neurotransmitters. Guidelines for use in children are not well established. In adults the initial dose is 2 to 4 mg. administered at 4 hour intervals and increased to 36 mg. as needed. It may cause drowsiness, nausea, hallucinations, and is hepatotoxic.

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A

Oral antispastic agents in CP

	Baclofen	*Diazepam*	*Dantrolene*
Mechanism	GABA analogue	Postsynaptic GABA-mimetic	Inhibits Ca^{++} release from sarcoplasmic reticulum
Dose	2.5 mg/day increased to 30 mg for 2 - 7 years 60 mg for 8 and above	0.12 - 0.8 mg/kg/day divided doses	0.5 mg/kg twice daily 3 mg/kg q.i.d.
Duration	2 - 6 hours	20 - 80 hours	4 - 15 hours
Side Effect	Seizure activity	Cognitive	Hepatotoxicity

Neuromuscular Blocking Agents
Local Anesthetics, Phenol, Botulinum Toxin

Consider using local anesthetics, alcohol, phenol and botulinum toxin as neuromuscular blocking agents [A] when treating focal spasticity.

Local anesthetics

Mechanism of effect

Local anesthetics block nerve conduction by changing membrane permeability to sodium ions. They affect both sensory and motor function in the area innervated by the nerve. This effect is completely reversible and causes no structural damage to the nerve. The effect starts within 3-15 minutes after the injection and lasts from 45 minutes to 8-12 hours depending on the type of drug used. Median nerve in the upper extremity and many nerves in the lower extremity are available for local anesthetic blocks [B].

Dosing and administration

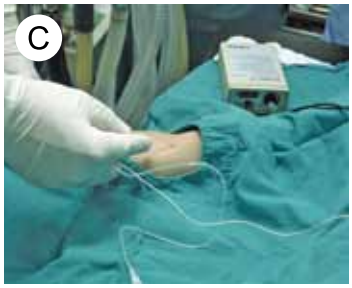
Lidocaine, etidocaine and bupivacaine are used for nerve blocks. Prefer bupivacaine because it is more potent and its duration of action is longer. It can be injected in amounts up to 3 mg/kg of 0.25 to 0.75% of a solution. Do a perineural injection when you want to block the motor, sensory and autonomic fibers in the nerve. A motor point block affects the motor fibers only.

A peripheral nerve stimulator that gives a low intensity electrical current through a needle electrode is used for blocks [C]. Use small needles and give short-lasting stimuli to localize the nerve more accurately. This makes the procedure less painful [D].

Local anesthetic blocks

- Median block
- Tibial block
- Obturator
- Femoral
- Sciatic

B



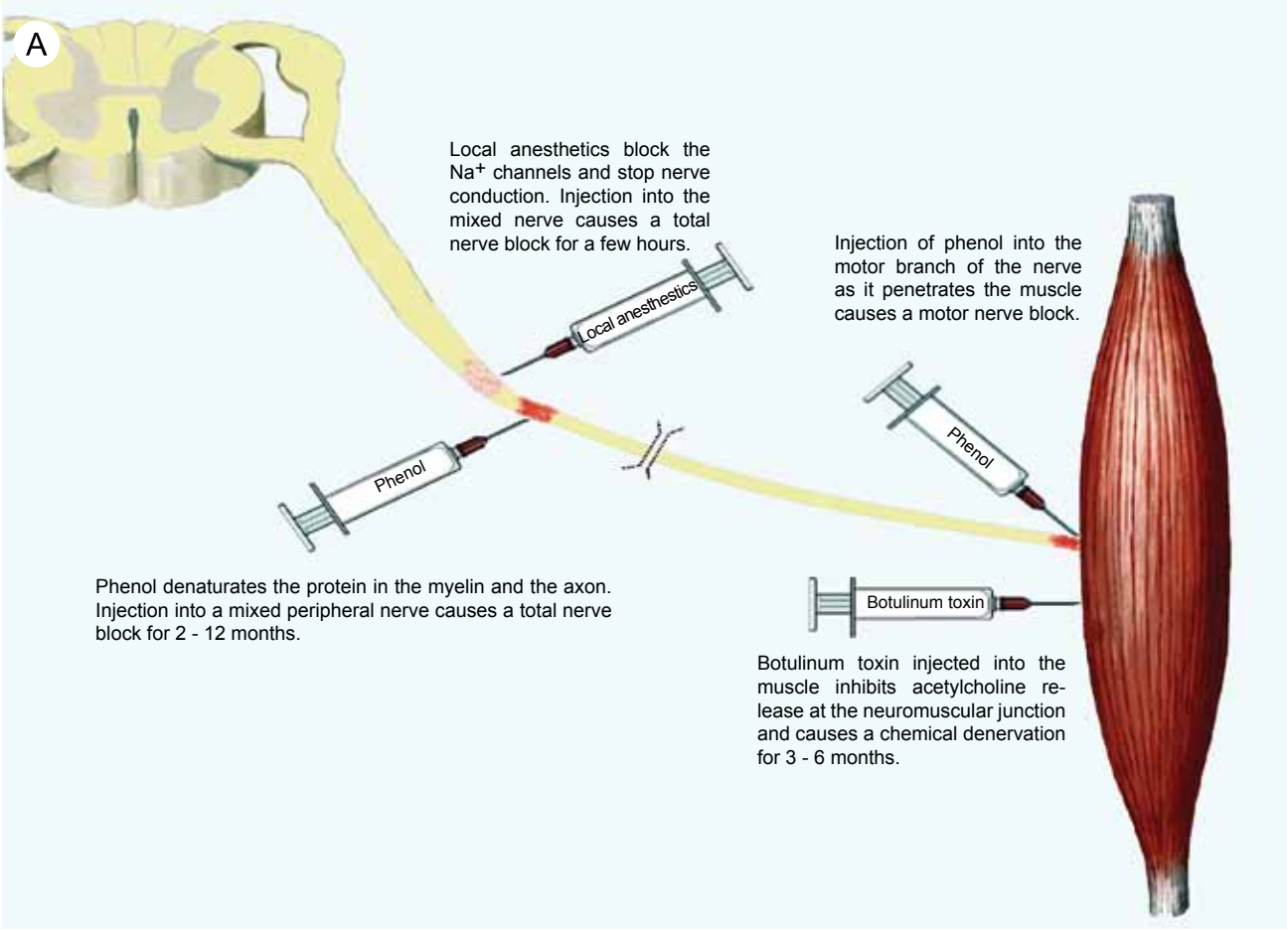
C

Electrical stimulation is used to locate the median nerve. Local anesthetic block to the median nerve results in total sensory and motor loss in the area innervated by the nerve. The effect lasts for a couple of hours.

D

Electrical stimulation technique

1. Locate the motor point or the nerve with the help of a stimulator. Charts exist for the location of each nerve.
2. Cleanse the skin. Choose the injection site and start stimulating the nerve. Adjust stimulation intensity first to a maximum, when the muscles innervated by the nerve begin to twitch, lower the intensity to 0.2-0.5 miliamperes.
3. If the muscle is still contracting, aspirate first and then inject the local anesthetic or phenol until the muscle is silent.
4. Increase the stimulus intensity to control the block. If there is no contraction at maximum stimulus intensity, the block is efficient. If not, inject more until the contraction stops.



A Indications for local anesthetic blocks
Differentiate spasticity from contracture
Predict functional changes
Distinguish the muscles that contribute to spasticity
Evaluate the presence of selective motor control

B Advantages of local anesthetic blocks
Reversible short duration effect
Relatively painless
Helps differentiate contracture from spasticity
Unmasks activity in the antagonists by relaxing the spastic muscles.

C Side effects and precautions
Hypersensitivity reaction
Hematoma at injection site
Sudden weakness may cause injuries in the unprepared patient
Systemic toxicity (dose related)

The injection may be painful and is best performed under general anesthesia in young children.

E Phenol blocks for lower extremity spasticity
The rectus femoris motor point block
The hamstring motor point block
Adductor muscle motor point block
Tibial nerve block

Indications

Local anesthetic blocks may be used as a diagnostic tool to differentiate spasticity from contracture and to predict functional changes with long term therapy [A]. The block may clarify which muscles contribute to spasticity and unmask selective motor control in the antagonist muscles if there is any. Block the median nerve at the elbow to evaluate the upper extremity. If flexion in the wrist and fingers is due to spasticity, the hand will relax completely a couple of minutes after the injection. Bring the fingers into extension while holding the wrist in extension. The joint will not relax if there is a contracture. Thus, a local anesthetic block aids the physician in the decision making process of treatment of the spastic hand.

Advantages

Local anesthetics have a short and reversible effect, so they are useful for diagnosis of the problem and differentiating contracture from dynamic spasticity [B].

Side effects and precautions

Local anesthetics rarely cause a hypersensitivity reaction in the form of a mild rash. Fatal anaphylactoid reactions have been reported. Hematoma may occur at the injection site. There can be significant changes in walking and transfers after a nerve block. Sudden decrease in muscle tone may result in falls and injuries in the early hours after the block. In high doses, local anesthetics may have systemic toxic side effects if they enter the systemic circulation by mistake. This is uncommon in children and in doses used for peripheral nerve blocks [C].

Chemical neurolysis: alcohol and phenol

Alcohol and phenol are chemical agents that block nerve conduction by creating a lesion in a portion of the nerve.

Alcohol

Ethyl alcohol acts as a local anesthetic by decreasing sodium and potassium conductance at the nerve membrane at low concentrations. It causes protein denaturation at higher concentrations such as 50%. Intramuscular injection of ethyl alcohol causes burning pain, therefore children must be injected under general anesthesia [D].

Even though alcohol has fewer adverse effects and is safer than phenol it has not been used as extensively in spasticity treatment possibly because of the pain it causes during the injection. Phenol blocks are generally used for lower extremity spasticity [E]. Recently botulinum toxin was added to the armamentarium of focal spasticity treatment [F].

F	Local Anesthetics	Phenol (6%)	Botulinum toxin A (Botox®)
Mechanism	Blocks sodium channels	Denatures protein	Inhibits acetylcholin release
Onset	Minutes	Less than an hour	Days
Duration	Hours	2-36 months	3-6 months
Dose	Bupivacaine (0.25-0.75%) <3mg/kg	Less than 10 ml (1 gm)	400 units at one single time
Precaution	Hypersensitivity	Pain-dysesthesia	None
Indication	Differentiate spasticity from contracture Test effects before long term blocks Relax muscles before casting	Proximal large muscles mainly motor nerves (no mixed nerve) More for hygiene and comfort In combination with BTX-A	All muscles accessible for injection Especially smaller muscles Active function Combination with phenol
Technique	Stimulation - motor point	Stimulation - motor point	Stimulation - EMG guide Motor point or end-plate targeting

Phenol

Mechanism of effect Phenol is benzyl-alcohol and has been used as a dysinfectant and antiseptic. It causes protein denaturation and non-selective tissue destruction in the injected area. Wallerian degeneration of neurons occurs in the weeks following injection. Most axons regrow over a period of time [A]. The effect of phenol starts rapidly because of its local anesthetic properties and lasts for up to 2 to 12 months.

Dosing and administration The usual dilution is 3 to 6% depending on the technique and the injection site. There are two techniques to apply phenol blocks: the motor point block and the motor nerve block. Motor point and motor nerve injection sites must be identified using electrical stimulation as explained in local anesthetic blocks. Electrically stimulating to find the motor points enables the physician to use very small quantities of the drug to obtain good clinical response [B].

Indications The advantages [C] include an early onset of action, longer duration of effect and low cost. In addition, there is no antibody formation to phenol so that larger, more powerful muscles may be treated without dosing considerations. Although the injection is painful at first, pain resolves in seconds because of its analgesic effects and injections are as easy as botulinum toxin injections for the experienced physician.

Side effects and precautions The main risks to be aware of when using phenol for spasticity management are permanent nerve injury, causalgia or neuropathic pain because of sensory fiber damage, tissue edema, venous thrombosis, and compartment syndrome resulting from large amounts of phenol in constrained space [B].

Avoid using phenol in the upper extremity because nerves in the upper limb are mainly mixed nerves and motor point blocks are difficult. Risks of dysesthesia, causalgia, venous thrombosis, and compartment syndromes are higher. Phenol is destructive to tissues, intramuscular administration in the small child may lead to unwanted and irreversible muscle fiber atrophy.

Combination treatment At present phenol has a rather small but useful place in spasticity treatment [D]. State-of-the-art treatment for focal spasticity relief is botulinum toxin. However, there is an upper limit to the amount of botulinum toxin that can be used in a single setting so a combination of phenol with botulinum toxin is preferred to better control multisegmental focal spasticity and to provide a longer duration of effect. Use phenol for large lower extremity muscles and botulinum toxin for smaller lower and all upper extremity muscles for multilevel injections whenever the necessary botulinum toxin dose exceeds the maximum amount you can use.

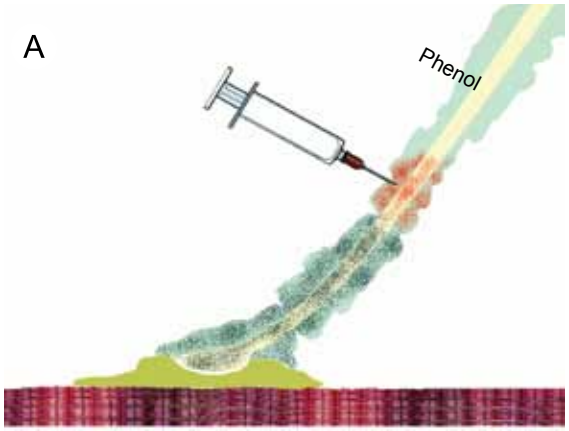
Botulinum toxin

Botulinum toxin, produced by the anaerobic bacteria Clostridium botulinum, is one of the most potent poisons known to man. In the past two decades it has been transformed into one of the most useful antispastic agents. Of the seven distinct toxins from A to G, only type A and B are used for therapeutic purposes. The structure of all toxins and their mechanism of action are similar, only their site of action is different.

The mechanism of effect

The toxin inhibits acetylcholine release at the neuromuscular junction causing a reversible chemodenervation. Studies

A

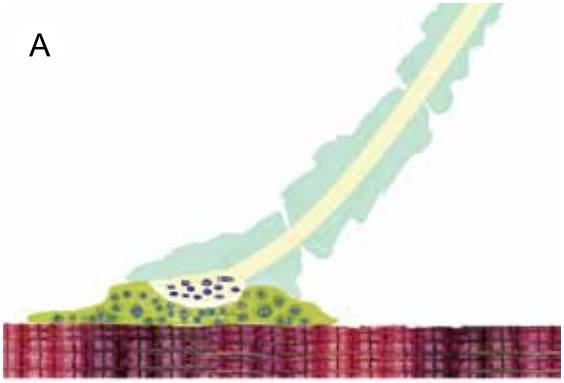


Phenol denaturates proteins in the myelin sheath and the axon. It also causes nonselective tissue destruction. The effects are reversible, most axons regrow.

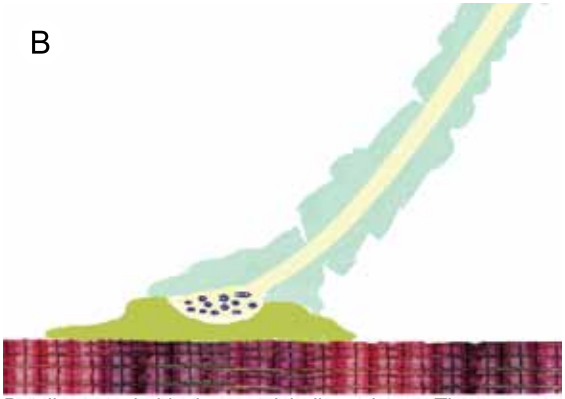
Advantages of phenol	B
Rapid action	
Longer duration	
Low cost	
No antibody formation	

Dysadvantages and precautions	C
Relatively painful injection	
Chronic dysesthesia and pain	
Peripheral edema, deep venous thrombosis	
Reversible sensory loss	
Systemic side effects (dose related)	
Relatively difficult technique	

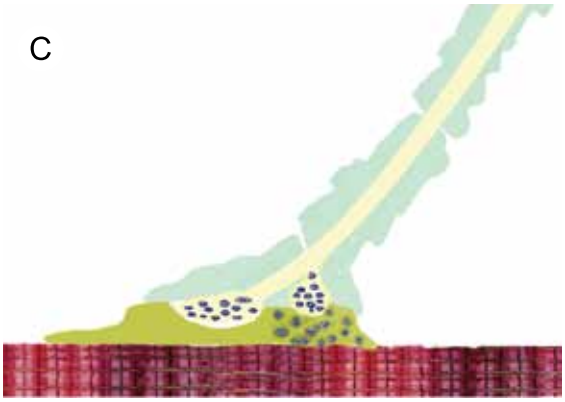
Hints on using phenol	D
Avoid using in the upper extremity	
Do not inject mixed peripheral nerves	
Only inject motor nerves	
The most common uses are rectus femoris motor point block obturator nerve block hamstring motor point block tibialis posterior nerve block (mixed nerve!) gastrocnemius motor point block	
Use 6 % concentration of phenol	
Maximum dose 1 ml/kg body weight	
The effects are immediately obvious	
Use 0.5-1 ml for motor point blocks Use up to 3 ml for nerve blocks	



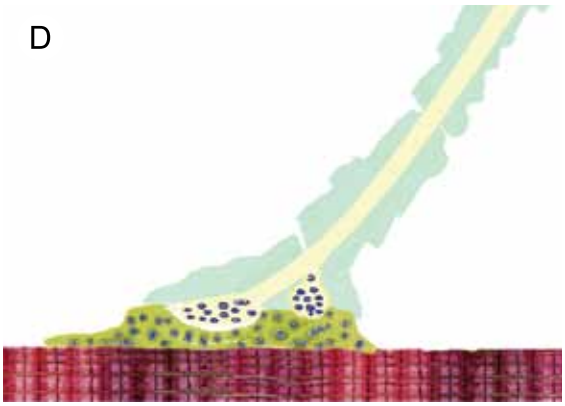
Muscle contraction depends on acetylcholine release from the axon terminal into the synaptic cleft.



Botulinum toxin blocks acetylcholine release. The axon terminal has normal amount of acetylcholine but the end-plate is not functional.



A nerve sprout forms immediately proximal to the dysfunctional end-plate and innervates the muscle fiber.



Eventually the original end-plate regains function as the effect of toxin wears off.

suggest that the toxin affects the muscle spindle and afferent nerve fibers as secondary actions.

Effect at the neuromuscular junction The toxin must enter the nerve endings to exert its effect. It becomes fully active once inside the cholinergic nerve terminal.

When the impulse for contraction arrives at the axon terminal acetylcholine (Ach) vesicles fuse with the nerve membrane and the Ach is released into the synaptic cleft. This causes excitation in the muscle fiber and muscle contraction [A]. The various serotypes of botulinum toxin act on different portions of the acetylcholine vesicle complex. Botulinum toxin inhibits the fusion of acetylcholine vesicles at the pre-synaptic membrane. Ach cannot be released into the synaptic cleft, the impulse from the nerve to the muscle fiber is blocked and the muscle fibers innervated by that axon cannot contract. This is chemical denervation [B]. The extent of muscle weakness created by the botulinum toxin depends on the serotype, dose and volume of toxin used.

The effect of botulinum toxin is reversible. Nerve sprouts form at the unmyelinated terminal axon immediately proximal to the end plate. These sprouts innervate the muscle fiber [C]. Eventually, the original neuromuscular junction regains function [D]. This terminates the clinical effect in 3 - 6 months and spasticity reappears.

Afferent effect The toxin may block the sensory afferents from the muscle spindle. This reduces spindle sensitivity and consequent reflex action.

Analgesic effect There is an analgesic effect of the toxin explained by a couple of mechanisms. First, decreasing spasticity decreases pain. Second, botulinum toxin affects afferent transmission and inhibits the release of substance P. Substance P is the primary mediator of pain in the spinal cord and the brain. Inhibition of its release together with the block in afferent transmission result in pain relief.

Specific pharmacology

The potency of the toxin is defined by mouse units. One mouse unit is the amount required to kill 50% of a group of female Swiss-Webster mice. There are two different commercial preparations for botulinum toxin; Botox® (Allergan), and Dysport® (Speywood) [B]. BTX-B is available as Myobloc™ in the United States and NeuroBloc® in Europe and elsewhere.

There are 100 units of botulinum toxin in one vial of Botox and 500 units in one vial of Dysport. The clinical potency of Botox and Dysport are influenced by numerous factors including the way they are produced. Therefore, the units are not interchangeable and there is no equivalence ratio between the two product.

Indications

Botulinum toxin injections have been used as a safe and effective treatment for spastic CP for the past 10 years. Botulinum toxin B is also becoming commercially available.

Botulinum toxin A is useful for overcoming activity limitations caused by spasticity; and is typically used in conjunction with orthoses and physical and/or occupational therapy. It may improve gait, limit progression of fixed deformity during growth, reduce intolerance to orthoses resulting from spasticity, and may delay the need for orthopaedic surgery in patients with GMFCS levels I, II & III.

The general indication for botulinum toxin injections in CP is 'the presence of a dynamic contracture, interfering with function, in the absence of a fixed muscular contracture'. If botulinum toxin injections are started at an early age and repeated as necessary, they can help prevent the development of muscle contractures and bony deformities. This helps to delay orthopaedic surgery until the gait is mature. The need for extensive surgical procedures may be eliminated if bony deformities are prevented by botulinum toxin.

The success of botulinum toxin administration depends on many factors. Patient selection is critical [A]. Children with spasticity who do not have fixed contractures benefit a great deal from treatment whereas patients with dyskinesia have a variable response and athetoids do not benefit at all.

The timing of the injections is controversial. Most clinicians agree that the earlier the spasticity is reduced, the better the outcome. Botulinum toxin can be injected as early as 18 months of age. There is no upper age limit, however, once the muscle is shortened as occurs with age, the effect of spasticity relief will not be apparent because of contracture.

Dosing and administration

Botulinum toxin dosing depends on which preparation is used. Dysport dosing is different than Botox and there is no equivalence ratio between the two preparations in terms of clinical effect. The doses mentioned here refer to Botox injections [B,C]. The amount changes according to the number of muscles to be treated, prior response of the patient if there are any prior injections and functional goals.

The dose limits range from 2 units to 29 units/kg of body weight, most common range being between 10-20 units/kg of body weight. Avoid injecting more than 400 to 600 units of total dose at any one time, injecting more than 50 units at one injection site and exceeding 20 units per kilogram per muscle at any one time. If there is a need for more toxin because of multilevel involvement, combine treatment with phenol. Inject larger muscles with phenol and use botulinum toxin for more distal and smaller muscles [D].

Targeting the neuromuscular junction during the injection using electrical stimulation guide may result in more effect for less volume. Even though no serious complications have been reported, it is a good idea to apply high doses under general anesthesia in the operating theatre. Reduce the dose if the child is small and has atrophic muscles, if the treatment is going to be repeated for a number of times and if multiple muscles are being injected. Severely spastic and larger muscles should receive a larger dose whereas less spastic and small muscles receive a smaller dose [E].

The amount of toxin given to one muscle must be divided into more than two injection sites, depending on the dose. Put a safe distance between two injection sites with high doses. This increases the diffusion of the toxin in the muscle and prevents it from entering the systemic circulation. Divide the total dose per muscle over more sites as much as possible. For example, for a 20 kg child who has a very spastic gastrocnemius muscle, the dose should be 6 U/kg/muscle, 120 U total. This dose should be divided into 4 injection sites, 30 units per site in the muscle.

Specific goals for botulinum toxin A treatment A

To improve walking in the spastic diplegic and hemiplegic child
To minimise adductor tone in the child with early hip subluxation
To decrease the spasms and pain in the spastic-athetoid patients
To reduce tone in the psoas muscle in patients with back pain because of hyperlordosis
As a simulation for orthopedic surgery, to have a general idea of how the child will be when spasticity is reduced.

General guidelines for upper extremity spasticity B

Muscles injected	*Dose range*	*Number of sites*
Biceps	2	2-3
Pronator teres	1	1
Flexor carpi radialis	2	1
Flexor carpi ulnaris	2	1
Flexor digitorum superficialis	2	1-2
Flexor digitorum profundus	2	1-2
Flexor pollicis longus	0.5-1	1
Adductor pollicis	0.5-1	1

General guidelines for lower extremity spasticity C

Muscles injected	*Dose range (units/kg of bw)*	*Number of sites per muscle*
Iliopsoas	2	2
Quadriceps	3-6	4
Medial hamstrings	3-6	3-4
Lateral hamstrings	2-3	2
Adductors	3-6	2
Gastrocnemius	3-6	1-2
Soleus	2-3	1
Tibialis posterior	1-3	1
In general maximum of 50 U/site		

Botox® dose modifiers D

	Decrease dose if	*Increase dose if*
Patient weight	Low	High
Duration of therapy	Chronic	Acute
Muscle bulk	Very small	Very large
Number of muscles injected simultaneously	Many	Few
Ashworth score	Low	Very high
Concern about weakness	High	Low
Results of previous therapy	Too much weakness	Inadequate response

Table reproduced with permission from WE MOVE New York www.mdvu.org.

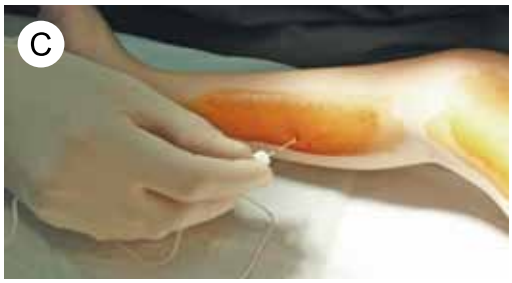
Recommended dosages E

Per muscle of lower limb	3-6 U/kg
Per kg total body weight:	12 U/kg proven dosage
Maximum dose per session	400 U
Frequency:	Not more than once every 3 months Usually at least 6 month intervals
Dilution	100 U in 1 or 2 ml 0.9% NaCl
Maximum dose per site	50 U

General guidelines for spastic CP A

Type of CP	*Muscles involved*	*Problem*
Hemiplegic	Rectus femoris	Stiff knee gait
	Gastrocsoleus & tibialis posterior	Pes equinovarus
	Flexor – pronator spasticity	Thumb in palm deformity, flexion of the wrist and digits
Diplegic	Multilevel lower extremity injections	Adductor - flexor spasticity of the hip
		Hamstring spasticity causing knee flexion
		Gastrocsoleus spasticity causing pes equinus
Quadriplegic	Hip adductors	Prevent hip subluxation
	Hamstring spasticity	Sacral sitting
		Sitting balance

Examine the child once again under general anesthesia. If there is no limitation of passive range of motion under general anesthesia, there are no contractures and the botulinum toxin injections will be useful. If there is limitation in joint motion indicating a fixed contracture, there will be a limited response to botulinum toxin.



Injection to the belly of the medial gastrocnemius with EMG guidance

Injecting the flexor pollicis brevis muscle using electrical stimulation

Patient Selection

Botulinum toxin is useful in various upper and lower extremity problems in spastic cerebral palsy cases [A].

Muscle selection

Choosing the right muscles to inject depends on a good clinical evaluation [B]. Evaluate passive range of motion at the ankle, knee and hip; measure spasticity using the modified Ashworth or the Tardieu scale and determine strength and selective motor control of different muscle groups of the lower limbs. Gait analysis using dynamic EMG may be helpful in complex cases.

Injection technique

Needle size depends on site of injection and physician preference. 1.0 ml tuberculin type syringes and 26-30 gauge, 1/2 inch (1.5 cm) needles are used. Teflon-coated monopolar injection needles are necessary for stimulation and injection with EMG or electrical stimulation guide [C].

Targeting Botulinum toxin dosing and injection technique is relatively easy. For optimal results the physicians must be experienced in managing children with CP. Difficult-to-localize muscles often require adjunctive methods to confirm injection sites and to target the region of the neuromuscular junctions. Electromyography (EMG), electrical stimulation [D], computerized tomography (CT), fluoroscopy, and ultrasound have been used to target the region of maximum muscle activity. The technique of electrical stimulation is the same as in local anesthetic blocks. Efficacy is maximal and adverse effects minimal if the muscles are targeted properly.

Sedation The injection is not painful, but may be a cause of distress in young children and in multilevel injections. It is rather difficult to inject certain muscles such as the hamstrings or iliopsoas in a fully awake and frightened child in the outpatient setting. Consider a simple sedative like diazepam or chloral hydrate when injecting single muscles in the outpatient clinic. Using EMG or ES guide and injecting multiple muscles is a considerable stress on the child so perform these under local anesthesia, conscious sedation using midazolam or general anesthesia.

Preparation Keep the toxin frozen in vial. Dilute with normal saline to the desired concentration prior to usage [E]. The toxin is in a vacuumed vial, when diluting hold the piston of the syringe steady because sudden inflow of saline into the vial may cause protein denaturation and loss of pharmacological activity. Then put a second needle through the lid to balance the negative pressure inside the vial before drawing back the diluted toxin.

Injection Clean the area, put sterile gloves on, localize the target muscle [A - L on next page], inject the desired amount into the muscle belly. You may need to inject at two or more sites depending on the dose and muscle size.

Dilutions E

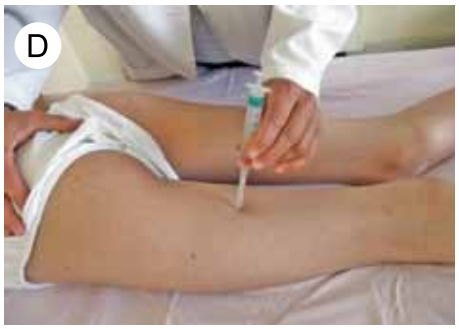
For 100 units of Botox preparation

Aimed final dilution	Saline added to vial
2.5 U/0.1 ml	4 ml.
5.0 U/0.1 ml	2 ml.
10.0 U/0.1 ml	1 ml.

Adductor longus muscle: Patient lies supine. Abduct the leg to 15°. Palpate the tendon arising from the pubic tubercle and insert the needle 2-4 finger breadths distal to the tubercle into the muscle belly.

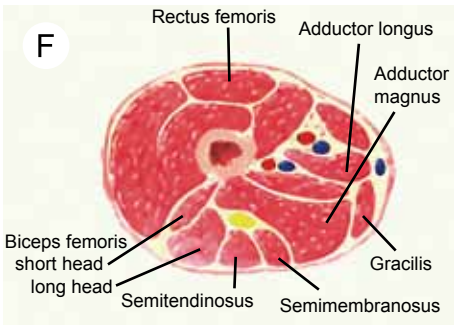
Adductor magnus muscle: Patient lies supine. Abduct and externally rotate the leg. Insert the needle midway between the medial femoral epicondyle and the pubic tubercle.

Rectus femoris: Patient lies supine. Insert the needle on the anterior aspect of the thigh, midway between the superior border of patella and the anterior superior iliac spine.



Medial hamstring muscles: Patient lies prone, Insert the needle at the midway on a line between the medial femoral epicondyle and the ischial tuberosity.

Lateral hamstring muscles: Patient lies prone. Insert the needle at the midway on a line between the fibula head and the ischial tuberosity.

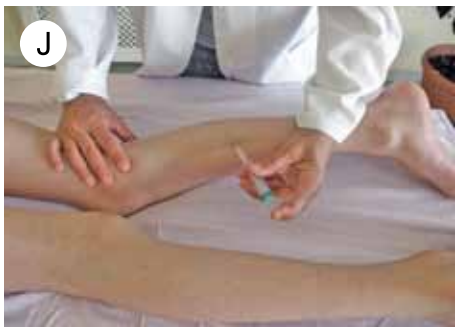


Coronal view of commonly injected thigh muscles

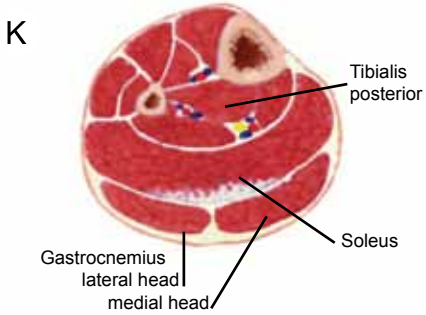
Gastrocnemius, medial head: Patient lies prone, leg extended. Insert at the most prominent point of the medial muscle mass (approximately 3 fingers to one handbreadth below the popliteal crease.)

Gastrocnemius, lateral head: Patient lies prone, leg extended. Insert at the most prominent point of the lateral muscle mass (approximately 3 fingers to one handbreadth below the popliteal crease.)

Soleus: Patient lies prone, the leg is extended. Insert the needle deep just distal to the belly of the gastrocnemius muscle, medial and anterior to the Achilles tendon.



Tibialis posterior: Patient lies prone, with the leg in internal rotation. Draw a line from the popliteal crease to the medial malleolus. Inject one finger breadth off the medial edge of the tibia, directly obliquely through the soleus and the flexor digitorum longus, just posterior to the tibia.



Coronal view of commonly injected calf muscles

Ultrasonographic guidance can be helpful especially when injecting deep muscles. Courtesy D. Ganjwala

Put the cast on and bivalve it if possible while the patient is still sedated so that the trauma of casting will be minimal.

Resistance	
Primary nonresponder	*Secondary non responder*
No response to initial injection	Relative or complete loss of effect after second injection
Presence of antibodies	Low dose Poor technique Change in spasticity Inappropriate reconstitution Inappropriate storage Antibody formation

B

Advantages	Dysadvantages
The minimality of side effects	Cost
Ease of application	Availability
Relatively painless injection	
Reversible effect (may not be an advantage)	
No permanent injury to tissues	

C

D Side effects
(Rare, all reversible)
Slight weakness at site of injection
Local pain
Flu-like syndrome
Generalized weakness
Incontinence

E Contraindications & precautions
Aminoglycoside antibiotic use
Pregnancy (for adult CP patients)
Lactation (for adult CP patients)

Post-injection treatment

The antispastic effect appears within 24 hours to 3 days after injection and becomes maximum at 10 days to a month. It lasts for 3 to 6 months. Some patients are golden responders in whom the antispastic effect lasts for over a year. Proper exercises, splinting and casting may increase the number of golden responders.

Casting for 2 to 3 weeks after injections may improve the results. Botulinum toxin relieves dynamic spasticity whereas casting addresses fixed contracture. Consider casting for two weeks beginning on the third day after the injection in severe cases. If injecting under conscious sedation or general anesthesia, put the cast on when the child is sedated or asleep [A].

Problems related to casting are psychological trauma of putting the cast on and taking it off and muscle atrophy.

Physical therapy Perform range of motion and strengthening exercises in an intensive manner to obtain maximum benefits from the injection. Intensive exercises and electrical stimulation after the injection may increase toxin uptake by the nerve terminal and potentiate the effect.

Orthotic management Continue bracing as prior. Brace tolerance generally increases after the injection.

Resistance

A small percent of children may not respond to initial injection of botulinum toxin. Consider one or more treatments before classifying patient as a "non-responder". A secondary non-responder is a child who shows a relative or complete loss of effect after a second injection. The reasons are too low a dose, poor injection technique, a change in the spastic muscles during treatment, inappropriate reconstitution or storage of toxin and the presence of neutralizing antibodies.

Development of resistance to botulinum toxin therapy is characterized by absence of any beneficial effect and by lack of muscle atrophy following the injection. Antitoxin antibodies are presumed responsible for most cases of resistance. Use the smallest possible effective dose and extend the time interval between treatments to at least 3 months to reduce the likelihood of antibody development. Botulinum toxin B or F may benefit those who have developed antibody resistance.

Advantages and dysadvantages

Side effects are few, mild and rare. The injection is relatively easy compared to phenol. There is no permanent tissue injury and all the effects are reversible. The cost is the only factor limiting toxin use [C].

Contraindications

Side effects are extremely few [D]. Slight weakness at injection site, local pain, fever, generalised weakness and fatigue presenting as a flu-like syndrome, respiratory tract infections, temporary incontinence and constipation have been reported with an incidence of 2-3%.

Contraindications include patients who are hypersensitive to any ingredient in botulinum toxin, who are using aminoglycoside antibiotics, pregnant or may become pregnant, or in lactation [E]. These contraindications are not absolute and not really relevant for children with CP. Patients who have a neuromuscular junction disease such as myasthenia like syndrome are not appropriate candidates for botulinum toxin therapy.

Conclusion

Botulinum toxin has an established place in the treatment of spasticity in cerebral palsy. Consider botulinum toxin treatment as early as two years of age and combine with other treatment options as the child grows older and spasticity begins to cause contractures and deformities [A].

The only factors limiting its use are high cost and restriction on the maximum dose per treatment session. The most common indications are young diplegic [B] and hemiplegic [C] children.

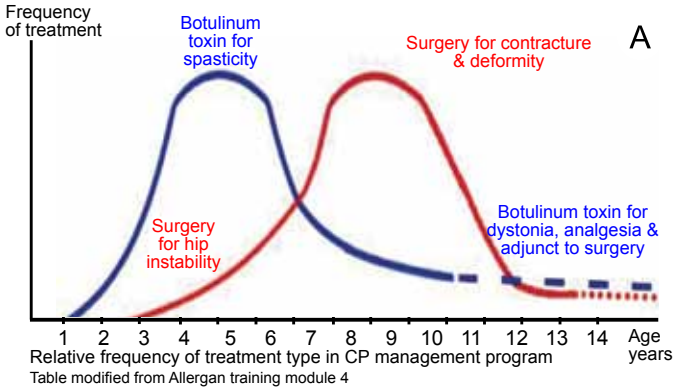
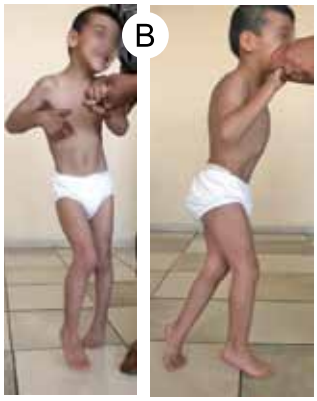
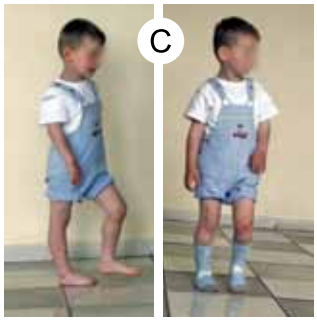


Table modified from Allergan training module 4



Diplegic 5 year old patient with jump gait, body weight 18 kilograms

Muscles to be injected	*Dose*	*Total dose per muscle*	*Number of injection site*
Right medial hamstring	4	72	2
Left medial hamstring	4	72	2
Right lateral hamstring	3	54	1
Left lateral hamstring	3	54	1
Right gastrocnemius	4	72	2
Left gastrocnemius	4	72	2
Total	396 units		10



Hemiplegic 4 year old patient with stiff knee and pes equinovarus, body weight 15 kilograms

Muscles to be injected	*Dose*	*Total dose per muscle*	*Number of injection sites*
Right quadriceps	4	60	4
Right gastrocnemius	5	75	2
Right tibialis posterior	2	30	1
Total	165 units		7

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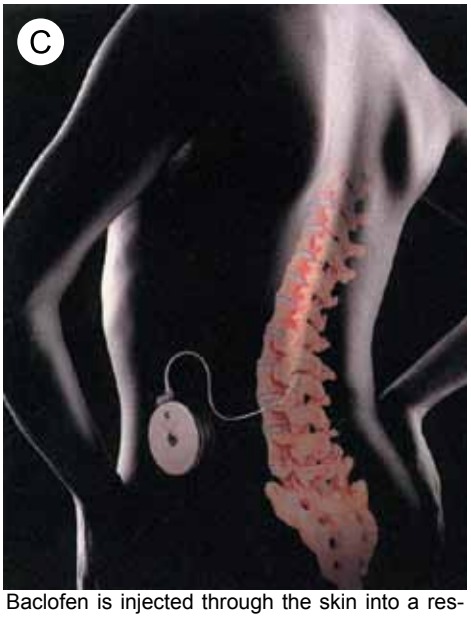
Indications (A)
Severe total body involved child
Severe dystonic or mixed CP
To ease burden of care
To enable sitting and transfers
To decrease spinal deformity
Diplegic children with severe spasticity interfering with ambulation

Before the implantation (B)
Answer these questions Is tone interefering with function ? Is tone interfering with patient care ?
Define type of involvement and clarify expected outcome
Evaluate family resources and cooperation
Evaluate the medical status of the child
Age
Is the abdomen large enough?
Is there recurrent infection?
Hydrocephalus?
Seizure activity ?
Evaluate financial resources
Perform test dose



Baclofen is injected through the skin into a reservoir placed in the abdominal wall. The reservoir also contains a programmable pump which is connected to the lumbar epidural space via a catheter. Courtesy of Medtronic Inc.

Intrathecal Baclofen (ITB)

Baclofen is one of the most potent antispastic drugs. It cannot easily cross the blood brain barrier because of its poor lipid solubility. This makes it difficult to reach therapeutic doses in the CNS. A novel method of introducing the baclofen directly into the CSF through an implantable pump and catheter system has been devised in the past decade and has become increasingly popular. Intrathecal administration enables the drug to reach the receptor site quicker with a much lesser side effect profile.

Indications for ITB

ITB is useful for the severely involved spastic, dystonic or mixed child [A]. The aim is to enable sitting in the wheelchair, make transfers easier, decrease spinal deformity, increase the comfort level and ease of care through a decrease in spasticity. ITB pumps have been used in severe spastic diplegia, but more research is needed before one can definitely recommend this form of therapy for this particular problem.

Factors to consider

Consider several factors before the implantation [B]. Look for spasticity interfering with function and patient care. Define the type of involvement and the expected outcome after the intervention. Family cooperation is absolutely essential because complications of ITB pumps are potentially life threatening. The pump can be inserted in cases above the age of three, with an abdomen large enough for implantation. Check for hydrocephalus. It should be under control if present, otherwise it increases the chance of CSF leak. Get appropriate medical treatment for seizure activity because baclofen decreases the seizure threshold. Examine the skin of the back. It must be intact, there must be no pressure sores or active infection anywhere in the body. Financial resources must be sufficient because both the implantation and maintenance cost a substantial amount.

Performing the test dose

After the initial decision to implant a baclofen pump, perform a test to evaluate the effect of the drug when given intrathecally. Introduce 50 micrograms of baclofen into the intrathecal space by bolus injection through a lumbar punction in the spastic total body involved child. Implant the pump if the child responds to this dose. If the child does not respond, use 75 to 100 micrograms in the consecutive trials on the following days. The effect of intrathecal baclofen starts at 1-2 hours after the injection, reaches a maximum at 4-6 hours and gradually diminishes after 8 hours. Perform the test with an intrathecal catheter placed at the level of the 9th thoracic vertebra for the dystonic child. Give a continuous infusion of baclofen. Children who show a decrease of one or more in the Ashworth scale for a six to eight hour period are good candidates for pump implantation.

Implanting the pump

A minor surgical procedure is necessary for pump implantation [C]. Introduce the catheter into the intrathecal space at the distal thoracic or lumbar spine. Push the catheter tip to upper thoracic levels in cases of upper extremity spasticity and dystonia. The catheter is attached to an externally programmable pump implanted into the abdominal wall. The pump is filled transcutaneously every 2-3 months depending on the dosing schedule.

Follow-up

Dosing and clinical evaluation

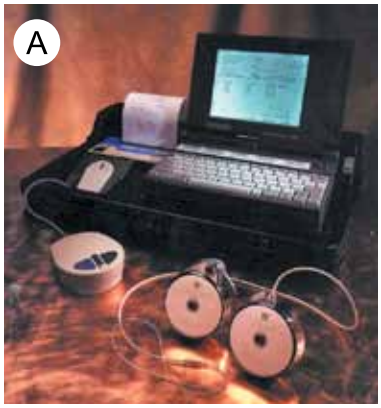
Intrathecal administration of baclofen provides a continuous infusion of the desired amount of baclofen into the CSF. A computer based remote control system makes it possible to regulate the daily dose [A]. The antispastic effects of intrathecal baclofen are obtained at 1% of the daily oral dose.

Begin with an initial daily dose of 25 micrograms and titrate up until there is a satisfactory reduction in spasticity. The dose is usually between 100 to 500 micrograms per day. A static dose is generally achieved within a year after implantation. The pump should be refilled at 1-3 month periods. Refills are made through a transcutaneous injection. The battery life of the pump is approximately 4-5 years.

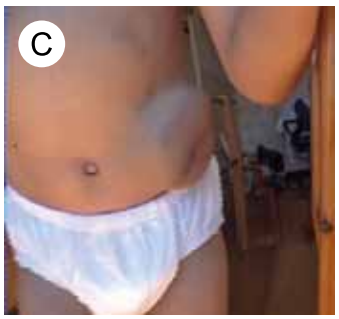
Begin an intensive physiotherapy program after pump implantation to reach functional goals [B,C]. Muscle weakness becomes prominent after a decrease in spasticity. Strengthening is important.

Complications

ITB pump implantation is expensive and the complication rate is moderately high. Complications include CNS infections, CSF leaks, and catheter related problems. Acute baclofen withdrawal syndrome [D] characterized by hallucinations, seizures, psychosis and rebound spasticity occurs if the baclofen flow to the CSF is interrupted. Signs of overdose are drowsiness, dizziness, somnolence, seizures, respiratory depression and loss of consciousness progressing to coma.



The intrathecal baclofen pump is remotely controlled by a computer. This enables the physician to increase or decrease the dose if necessary. Bolus injections may also be given.
Courtesy of Medtronic



The child's abdomen must be large enough for the pump. Sometimes the pump protrudes from under the skin and becomes vulnerable to trauma or infection.

D	Symptoms of acute baclofen withdrawal
	Acute increased tone
	Spasms
	Paresthesias
	Profuse sweating
	Dysphoria
	Hallucinations
	Seizures

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Selective Dorsal Rhizotomy and Other Neurosurgical Treatment Modalities

Selective dorsal rhizotomy (SDR) involves sectioning of the dorsal column rootlets to interrupt the spinal reflex arc [A]. This inhibits the afferent input from the muscle and tendons and reduces the efferent activity at the level of the spinal cord. The advantage of SDR is a global muscle tone reduction in lower extremities without producing weakness. All the lower extremity muscles are affected. The effects are permanent and weakness is not a major issue, however, there is loss of superficial and deep sensation.

Indications

Patient selection is important for success of the intervention. The ideal patient [B] is an independent ambulatory diplegic child between the ages of 3-10 with pure spasticity, no fixed contractures, good strength and balance with spasticity being the major limitation to function. Family commitment is essential for success because there is a need for long term intensive physiotherapy after the procedure. The extent of functional improvements cannot always be related to SDR itself because the patients also receive long and intensive hours of physiotherapy after the procedure for at least a year.

Technique

A laminectomy is done under general anesthesia and the posterior roots are exposed. EMG monitorization is recommended to determine which rootlets should be cut. The rootlets are stimulated electrically and the response from the muscles are observed. This way, the most active rootlets are localized. Up to 30-50% of the dorsal rootlets at each level from L2 to S1 are cut. In some centers, the L1 rootlets are also cut to assist in reduction of psoas activity. S2-S4 rootlets must be spared to preserve bladder function.

Follow-up

Expected results of the procedure are a loss of deep tendon reflexes, decrease in muscle tone, an improved gait pattern and smoothness of gait. Energy consumption may improve if walking is very inefficient prior to surgery. Sensory loss is usually transient though long term effects are not clear.

There is a need for extensive postoperative rehabilitation. After surgery, the therapy must focus on strengthening. Orthopaedic surgery is still necessary usually for foot instability (excessive valgus), rotational abnormalities and contractures. Continued gait improvements are minimal between 1 and 2 years after surgery.

Contraindications

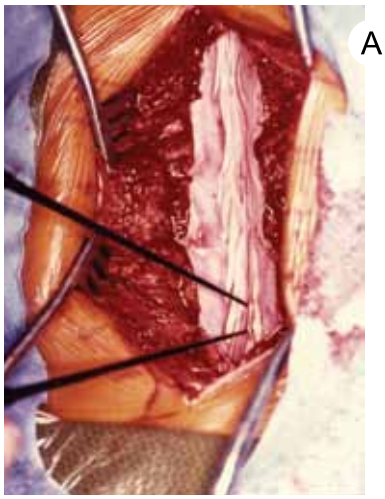
SDR is contraindicated in patients who have extrapyramidal findings, significant weakness or contractures, spinal abnormality and poor family support and commitment.

Side effects & Precautions

There are concerns regarding the development of hip instability and spinal deformity after SDR. Proprioceptive sensory loss is common and the long term effects are unknown.

Other neurosurgical treatment modalities

Deep brain stimulation and magnetic repetitive stimulation have all been tried in the CP patient with limited success [C]. Certain neurosurgical procedures such as thalamotomy and stereotaxic surgery have not produced satisfactory results.



Selective dorsal rhizotomy is technically difficult. The surgeon must be familiar with the anatomy of the spine and the spinal cord, must use electrophysiological monitoring to determine which and how many of the rootlets (*) he wants to cut and must be careful not to damage the cord in any way. The long term effects of SDR on joint integrity and muscle function are yet unknown.

B

The ideal SDR candidate
Diplegic child
Age 3-10
Independent ambulator
Pure spasticity
No fixed contractures
Good strength and balance
Reasonable selective motor control
Family commitment

C

Neurosurgical procedures in spasticity		
Procedure	*Target*	*Result*
Stereotaxic encephalotomy	Globus pallidus Ventrolateral thalamic nuclei	Variable-poor
Cerebellar stimulation	Cerebellum	Poor
Cervical rhizotomy	C1-C3	Variable-complications
Selective dorsal rhizotomy	L2-S2 selected rootlets	Variable-good
Neurectomy	Peripheral nerves	Variable, may cause chronic pain

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Hemiplegia

Hemiplegic children have involvement of the arm and leg on one side of the body [A]. The upper extremity is more severely involved than the lower [B]. Spastic hemiplegia constitutes 20% of cases with spastic CP. These children generally have very few associated problems. Communication is unimpaired most of the time. They may have seizures, learning and behavioural problems. Functional prognosis is good compared to other types because one side of the body is normal. All hemiplegic children learn to walk by the age of three. They become independent in the activities of daily living. Seizures, mild mental retardation, learning difficulties and behavioural disturbances may complicate the management and integration into the society.

Common musculoskeletal problems

The shoulder is adducted and internally rotated, the elbow is flexed and pronated, the wrist and fingers are flexed, the thumb is in the palm. The hip is flexed and internally rotated, the knee is flexed or extended, the ankle is in plantar flexion. The foot is generally in varus, although valgus deformity may also be seen. The hemiplegic side is short and atrophic depending on the severity of involvement [C].

Treatment consists of physiotherapy, occupational therapy, bracing, botulinum toxin injections and orthopaedic surgery [D]. Some children may need speech therapy and antiepileptic medication.

Physiotherapy & occupational therapy

Motor problems of the hemiplegic child are usually mild. Physiotherapy is prescribed to prevent contractures of the involved side, to strengthen the weak muscles, to enable functional use of the upper extremity and to establish a better walking pattern.

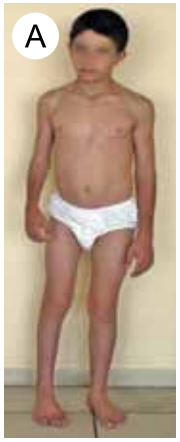
The basic program for the lower extremity consists of hip, knee, ankle range of motion exercises; rectus femoris, hamstring and gastrocnemius muscle stretching and agonist muscle strengthening. Do not neglect the back extensors and pelvic girdle muscles. Prescribe occupational therapy to gain hand function. Activities that involve both hands may improve the use of the involved side. Inhibiting the sound extremity and forcing the involved one to work is a novel method called constraint induced therapy. The main principle of constraint-induced movement therapy is restraint of the uninvolved hand and intensive practice with the involved hand, during a specified time period. This method has certain beneficial effects but it may be frustrating for most children.

Children with hemiplegia do not need physiotherapy for ambulation. Prognosis for independent walking is very good. Physiotherapy is beneficial to prevent contractures of the ankle. In most of the cases the physiotherapy and occupational therapy can be accomplished on an outpatient basis or home program.

Botulinum toxin A

Botulinum toxin injections are used for upper and lower extremity spasticity in the young child [E]. The toxin reduces gastrocnemius-soleus and rectus femoris spasticity in the lower extremity. The child uses his braces more efficiently and may develop a better walking pattern. Early relief of spasticity may prevent shortening of the gastrocnemius muscle and delay or eliminate the need for surgical intervention.

In the upper extremity, inject botulinum toxin to relax wrist, finger and thumb flexors so that the child may gain forearm supination and wrist stabilization.



Typical problems of the hemiplegic child consist of difficulty using the hand, walking on tiptoes and falling frequently.

Musculoskeletal problems in hemiplegia C

Upper extremity		*Lower extremity*	
Shoulder	Internal rotation Adduction	Hip	Flexion Internal rotation
Elbow	Pronation Flexion	Knee	Flexion Extension
Wrist	Flexion	Ankle	Plantar flexion
Hand	Flexion Thumb-in-palm	Foot	Varus

Treatment in hemiplegia D

Physiotherapy	Prevent contractures Strengthen weak muscles Establish a better walking pattern
Occupational therapy	Functional use of upper extremity Activities of daily living
Bracing	Lower extremity Solid or hinged AFOs
	Upper extremity Functional or resting hand splints
Botulinum toxin A	Lower extremity Rectus femoris and gastroc spasticity
	Upper extremity Pronator flexor spasticity
Orthopaedic surgery	Correction of Pes equinovarus Stiff knee Femoral anteversion

Botulinum toxin A injections in hemiplegia E

Location	*Problem*	*Muscle*	*Dose units/kg bw*
Upper extremity	Elbow flexion	Biceps	2
	Forearm pronation	Pronator teres	1
	Wrist flexion	Flexor carpi radialis	2
		Flexor carpi ulnaris	2
	Finger flexion	Flexor digitorum sup	2
		Flexor digitorum prof.	2
	Thumb in palm	Adductor pollicis	0,5
Lower extremity	Rectus femoris	Stiff knee	3-6
	Gastrocnemius	Pes equinus	3-6
	Tibialis posterior	Pes varus	1-3

Relaxing the spastic muscles with botulinum toxin injections may aid the treatment team to visualize how the child will function when his spastic muscles are surgically lengthened.However, the toxin cannot show its real effect in some older children with already shortened muscles.

Botulinum toxin may be combined with surgery in the older child. Inject muscles which have mild spasticity and no shortening with botulinum toxin; and surgically lengthen the severely spastic short muscles. This combination approach adopted in the recent years enables a swifter return of function, less complications and less muscle weakness because of less extensive orthopaedic surgery.

Bracing

Upper extremity bracing

There are two indications for hand splints in hemiplegia. One is to prevent deformity and the other is to improve function. Night splints help stretch muscles and maintain range of motion.Tone usually decreases at night, therefore the use of resting splints at night to prevent deformity is questionable. The child's compliance with night splints is generally poor.

Use day splints to increase function by either supporting the wrist in 10° extension, the thumb in opposition or both. Keep in mind that day splints prevent sensory input in the already compromised hand.

Lower extremity bracing [A]

AFOs stabilize the ankle and foot and keep it in the plantigrade position for weight bearing [B]. They are set in 5° dorsiflexion to avoid genu recurvatum or at neutral to prevent knee flexion. If the foot remains fixed the child has to extend the knee. Correct all fixed contractures before giving braces. Use hinged AFOs for mono and hemiplegic patients especially when they have active dorsiflexion.

Orthopaedic surgery

The usual indications for surgery are pes equinus, pes varus and stiff knee. Thumb-in-palm and wrist flexion deformity also respond to surgery. Perform soft tissue procedures around 5-6 years of age. Wait until at least 8 years of age for bone procedures unless the deformity is causing a functional problem.

Delay upper extremity surgery for function (age 6 to 12) until the child is mature enough to cooperate with postoperative rehabilitation.

The foot

The common problems of the foot in hemiplegia are pes equinus and varus. They often occur in combination with each other and with knee problems [C]. Evaluate the hips, knees and feet as a whole when examining the lower extremity.

Pes equinus

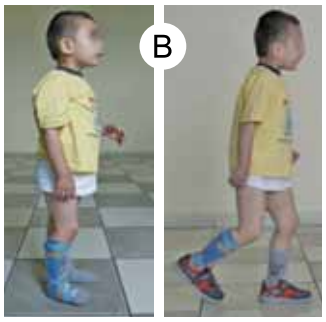
Pes equinus is ankle plantar flexion during gait [D] that occurs because of gastrocnemius-soleus spasticity. It may be dynamic or static. Dynamic equinus occurs only during walking secondary to gastrocnemius spasticity. Passive ankle dorsiflexion is not limited. When the gastrocnemius-solues muscle is short, passive ankle dorsiflexion is limited and static pes equinus occurs [E].

The child with pes equinus bears body weight on the metatarsals. Callosities occur in adolescents and adults. Step length is short, toe clearance in swing is inadequate and the ankle is unstable. Sometimes the discrepancy caused by pes equinus may result in pelvic obliquity [F].

Stretching and corrective casting Treat children younger than age 5 with stretching exercises and corrective casting. Apply corrective casts for 2 or more consecutive sessions for 3 weeks in dynamic and mild static contractures. Consider injecting botulinum toxin prior to casting to improve the results. Always prescribe stretching exercises and plastic AFOs after casting.

Type of brace	Indication
Solid AFO	Equinus and equinovarus
AFO in 5° dorsiflexion	Equinus & genu recurvatum
Hinged AFOs	Equinus and equinovarus if the child: can tolerate the hinge has varus-valgus control has 5° passive dorsiflexion
Supramallleolar orthoses (SMO)	Mild varus - valgus deformity without equinus

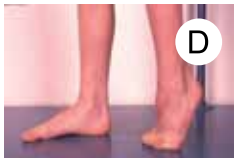
A



The AFO provides stability in stance and foot clearance in swing.

The foot in hemiplegia
Pes equinus, pes varus & combination
In isolation or with knee & hip problems
Watch the child walk
Examine passive range of motion
Test for flexibility
Examine active range of motion
Gait analysis & pedobarography

C



Neglected gastrocnemius spasticity results in fixed pes equinus deformity.

Causes of pes equinus
Gastrocnemius spasticity
Soleus spasticity

E

Pes equinus results in
Inadequate toe clearance during swing
Instability in stance
Short step length
Callosities
Difficulty with shoewear

F

Make sure that the corrective force is applied at the ankle joint during casting. If the cast does not fit properly, the force stresses the midfoot and causes rocker bottom deformity.

Botulinum toxin Botulinum toxin is the treatment of choice in very young children with gastrocnemius spasticity, recommended as a time-buying agent in children who are not suitable for surgery. Inject botulinum toxin into the spastic gastrocnemius muscle. Do not exceed 50 units per injection site. Apply a cast or use a full time solid AFO after the injection to improve and lengthen the effect.

Surgical treatment Consider surgical treatment in children who have walking difficulty because of a dynamic or static contracture [A]. Lengthen the gastrocnemius muscle by selectively incising its tendon through a full thickness transverse cut at the musculoskeletal junction as it combines with the soleus. Warren -White or Hoke are two different techniques advised to perform this operation. Lengthen the Achilles tendon if there is soleus contracture as well. Cut the Achilles tendon percutaneously by multiple partial tendon incisions and then dorsiflex the ankle with the knee in extension to allow the cut portions to slide in place. Try Z-lengthening in older and neglected cases where the tendon is markedly short.

Try and gain at least 15-20° dorsiflexion at the ankle. Put the child in a short leg cast with 5° dorsiflexion. Never cast in excessive dorsiflexion.

Postoperative care Keep the child in a short leg cast for 3 weeks. Use the cast up to 6 weeks for older children or after Z- lengthening. Begin ambulation as early as 2-3 days after surgery. Allow full weight bearing with crutches. Put the patient in AFOs right after cast removal and have him wear it night and day. Discard the brace during the day and use it as a night splint only after 3 months in children with good voluntary tibialis anterior function. Recurrence is high in patients with no voluntary tibialis anterior function. They must use their brace until they gain active dorsiflexion [B]. A pedobarography is useful to evaluate the outcome [C].

Complications of pes equinus surgery are rare [D]. There is a 25% risk of recurrence because of weakness of tibialis anterior muscle and also skeletal growth. Recurrence risk increases in cases who have inadequate lengthening or do not wear braces [E]. Patients younger than 5 years of age have a high risk of recurrence. Excessive lengthening of the triceps surae causes pes calcaneus deformity and the push-off is weakened.

Pes varus

Pes varus is characterized by increased inversion and exaggerated weight bearing on the lateral margin of the foot [F]. The causes are tibialis anterior, tibialis posterior and triceps surae spasticity with peroneal muscle weakness [G]. The more common tibialis posterior spasticity causes hindfoot varus, and tibialis anterior spasticity causes midfoot varus. Pes equinus usually accompanies pes varus, pure varus is relatively rare. The hemiplegic child with increased femoral anteversion or internal tibial torsion has intoeing gait that looks like varus.

Varus over 10° causes problems with foot clearance during swing and stability in stance. Older children have difficulty wearing shoes. Callosities form under the fifth metatarsal [H].

Stretching and corrective casting Treat flexible pes varus with stretching exercises and braces. Inject botulinum toxin to the spastic tibialis posterior to decrease spasticity and achieve foot alignment with a brace. Perform the injection with EMG or electrical stimulation guide to localize the deep lying tibialis posterior muscle.

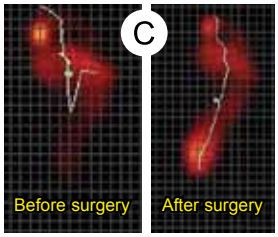
A Surgical options for pes equinus

Silfverskiöld test negative	Gastrocnemius lengthening
Silfverskiöld test positive	Achilles tendon lengthening
Severe neglected equinus	Posterior capsulotomy*

** Combine with Achilles tendon lengthening*

B Postoperative care: Pes equinus surgery

Cast	3 weeks in young child 6 weeks in older child
Ambulation	2 - 3 days
Weight bearing	Full
Brace	Until active dorsiflexion appears



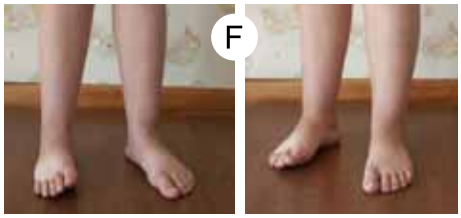
Pedobarography shows the pressure distribution of the foot and is useful in evaluating pes equinus.

D Complications of pes equinus surgery

- Recurrence
- Excessive lengthening
- Pressure sores

Causes of recurrence after pes equinus surgery (E)

- Weak tibialis anterior
- Musculoskeletal growth
- Inadequate lengthening
- Noncompliance with the brace
- Early surgery (age < 5 years)
- Z - lengthening



The muscle responsible for pes varus may be difficult to determine on physical examination.

Causes of the varus foot (G)

Tibialis posterior spasticity	Hindfoot varus
Tibialis anterior spasticity	Midfoot varus
Triceps surae spasticity	Ankle varus

H Pes varus results in

- Poor foot clearance during swing
- Instability in stance
- Difficulty with shoewear
- Painful callosities
- Cosmetic problems

Inject the gastrocnemius and soleus simultaneously. Varus deformity tends to worsen after 5-6 years of age in many patients. Consider surgical treatment if the deformity becomes fixed.

Surgical treatment Correct muscle imbalance in young children before bony deformities develop. The choice of surgical method depends on the involved muscle [A].

The tiptoe test is a good method to evaluate the posterior tibialis muscle. Ask the child to walk on his toes. Because the tibialis anterior does not contract during tiptoe walking, persistence of varus shows spasticity of the tibialis posterior muscle.

Another method of evaluating these two muscles is dynamic EMG. However, EMG needles inserted into the muscles disturb the child's gait and therefore cannot be useful.

Pedobarography may also help determine the true cause of equinovarus [B]. Overactivity of the tibialis posterior will cause more weight bearing on the fifth metatarsal whereas overactivity of the tibialis anterior will cause a cavus weight bearing pattern with increased pressure over the first and fifth metatarsals.

Soft tissue surgery Lengthen the tibialis posterior muscle at the musculotendinous junction and perform a split transfer of the tibialis anterior tendon (SPLATT). Do a split transfer of the posterior tibialis tendon (SPLOTT) if the tibialis anterior muscle is weak, or when there is posterior tibialis contraction during swing. This operation preserves plantar flexion force and replaces weak peroneals. Results may not be optimal though recurrence is rare. Combine triceps lengthening with other soft tissue surgeries if the triceps muscle is short [C].

Bone surgery There is a need for bone surgery in children with bony deformity. Wait until the child is 7 - 8 years old for a calcaneal osteotomy. Combine calcaneal osteotomy with tendon surgery to achieve satisfactory correction. Triple arthrodesis is an option for severe deformities in older children. Do not perform triple arthrodesis before 15 years of age. Postoperative care is similar to pes equinus.

The knee

Common knee problems in hemiplegia are flexed knee, genu recurvatum and stiff knee [D].

Flexed knee

The predominant pattern in hemiplegia is the flexed knee that is usually associated with triceps and hamstring spasticity. Use an AFO for mild cases, combine with botulinum toxin injections to the hamstrings if necessary. In older children and in severe cases lengthen the hamstrings surgically.

Genu recurvatum

Genu recurvatum is defined as knee hyperextension during stance. It occurs secondary to pes equinus, spasticity of rectus femoris, hamstring weakness or their combinations [D].

Conservative treatment Consider botulinum toxin injection to rectus femoris and gastrocnemius-soleus muscles. A plastic hinged or solid AFO with plantar flexion stop set at 5° - 7° dorsiflexion may prevent genu recurvatum [E].

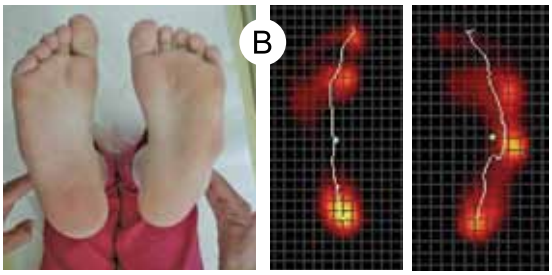
Surgical treatment Depending on the etiology, lengthen the triceps surae and/or the rectus femoris. Rectus femoris transfer to medial hamstrings is another option [F].

Stiff knee

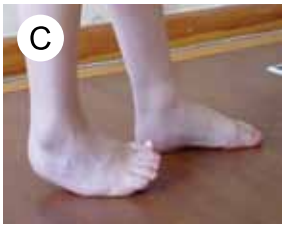
Stiff knee gait is defined as decreased knee flexion (less than 30°) during the gait cycle. The cause of stiff knee gait is rectus femoris spasticity. The spastic rectus femoris contracts during the swing phase and prevents the knee from going into flexion. Treatment is often difficult. Try botulinum toxin injections to the spastic rectus femoris. Lengthen or transfer the rectus femoris to the medial hamstring if necessary.

A Surgical options for pes varus

*Split tibialis anterior muscle transfer
*Tibialis posterior lengthening
Split tibialis posterior muscle transfer
Achilles tendon lengthening
Calcaneal osteotomy
Triple arthrodesis
** These two operations are usually combined.*



Pure varus deformity results in excessive weight bearing on the lateral margin of the foot. The increased load on the lateral aspect of the foot can be detected by pedobarography.



Pes varus and pes equinus frequently occur together.

The knee in hemiplegia D

Problem	*Definition*	*Cause*
Flexed knee	Flexion in stance	Hamstring spasticity
Genu recurvatum	Hyperextension in stance	Rectus femoris spasticity Pes equinus Hamstring weakness
Stiff knee	Decreased flexion during gait	Rectus femoris spasticity

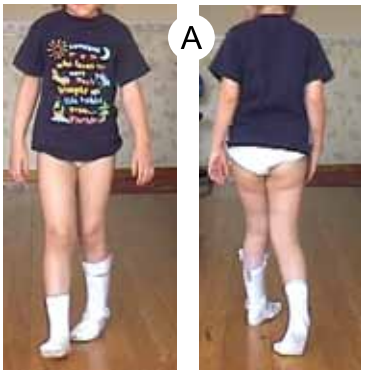
Genu recurvatum is generally secondary to pes equinus. Rectus femoris spasticity contributes to the problem.

F Treatment of genu recurvatum

Pes equinus	Gastrocnemius lengthening
Rectus femoris spasticity	Rectus femoris lengthening or transfer

The hip

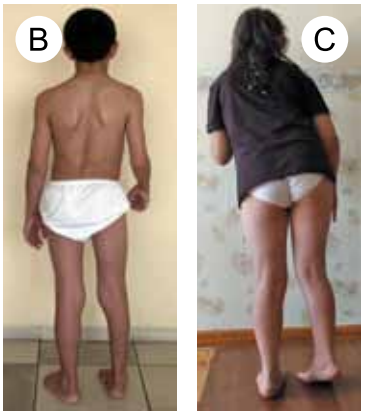
Hip problems are not common in hemiplegic children. Hip subluxation is extremely rare. Some children have a flexion-adduction and internal rotation deformity. Persistent femoral anteversion causes hip internal rotation and intoeing gait [A]. Internal rotation of the extremity disturbs foot clearance, the child may trip over his foot and fall. Children with intoeing develop a compensatory dynamic equinus that can be mistaken for gastrocnemius spasticity. Consider lengthening the iliopsoas and adductor muscles and performing proximal or distal femoral rotation osteotomies according to the patients' needs. Correct a compensatory tibial external rotation with a distal tibial osteotomy.



Femoral anteversion causes hip internal rotation and intoeing gait.

Limb length discrepancy

Almost all hemiplegic children have slight atrophy and shortening of the involved lower extremity [B,C]. The discrepancy is generally less than 15 mms. Shoe inserts or surgery are not necessary. On the contrary, having a slightly shorter leg on the involved side helps toe clearance during swing. Consider a shoe insert in a discrepancy of over 15 mm to prevent pelvic obliquity.



Limb length discrepancy is common, may cause pelvic obliquity and secondary scoliosis.

Management of hemiplegic gait

There are four types of hemiplegic gait [D].

Type 1: There is weakness of the tibialis anterior and an adequate gastrocnemius-soleus length. The child shows foot drop in the swing phase. Use a hinged AFO allowing free dorsiflexion.

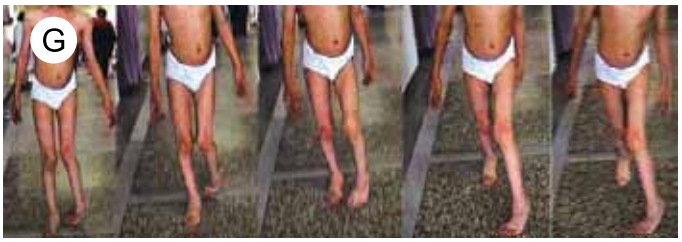
Type 2: Gastrocnemius-soleus muscle is short in addition to tibialis anterior weakness. The child compensates with knee hyperextension in midstance [E]. Inject botulinum toxin to the gastrocnemius-soleus complex if the deformity is dynamic. If static, serial casting or surgery are options. Use hinged AFOs after surgery.

Type 3: There is persistent knee flexion in stance phase and decreased knee motion in swing phase in addition to the above findings. This is defined as stiff knee gait. The treatment should include hamstring lengthenings to treat knee flexion if they are active during swing as well as rectus femoris transfers to semitendinosus to treat decreased knee motion in swing.

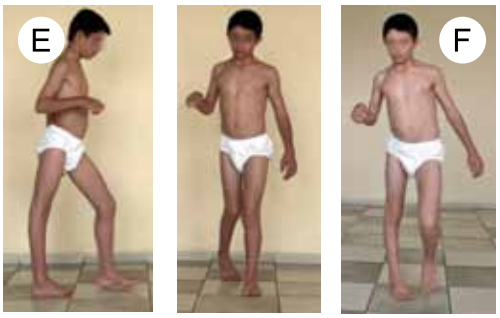
Type 4: There is adduction and flexion of the hip in addition to the findings above [F]. Lengthen the hip adductors and flexors if necessary. Bony deformities such as excessive internal femoral rotation and tibial torsion may also be seen. Treat bony deformities with appropriate rotational osteotomies [G].

Hemiplegic gait (According to Winters & Gage) D

Type	*Problem*	*Result*	*Treatment*
I	Weak tibialis anterior Adequate gastroc-soleus	Foot drop in swing	Hinged AFO Allowing free dorsiflexion
II	Weak tibialis anterior Short gastroc-soleus	Foot drop in swing Genu recurvatum	Botulinum toxin to gastrocnemius Serial casting Surgery Hinged AFOs
III	In addition to above: Persistent knee flexion Decreased knee motion in swing	In addition to above: Stiff knee Knee flexion	In addition to above: Lengthen hamstrings Transfer rectus femoris to semitendi-nosus
IV	In addition to above: Hip adduction, flexion & internal femoral rotation	In addition to above: Intoeing	In addition to above: Release at the hip Derotation osteotomy



Rotational osteotomies maybe necessary to correct the excessive internal rotation in hemiplegia.



Hemiplegic gait is characterised by pes equinus, genu recurvatum, internal femoral rotation and hip adduction.

Upper extremity

Lack of voluntary control, sensory impairment, muscular imbalance caused by spasticity and weakness, joint contractures, and articular instability all contribute to the upper extremity problem in CP [A]. The child has difficulty using the hand. The shoulder is in internal rotation and adduction, elbow in flexion, forearm in pronation, wrist in flexion and ulnar deviation, and thumb in adduction and flexion (thumb-in-palm) [B]. The child cannot position the hand in space, grasping an object and letting go are difficult [C,D]. Since children with hemiplegia have a normal upper extremity that they use in daily life, these deformities do not compromise the activities of daily living substantially. Hemiplegic children tend to ignore the plegic side due to sensory deficits. This neglect reinforces the impairment, inhibits the development of hand-eye coordination and prevents function in the involved extremity. The child cannot learn to use his involved hand even if he has the potential.

The aim of treatment is to increase function, improve hygiene and cosmesis. The hand is a tool also for social communication. Even minor improvements in hand cosmesis increase the patient's self esteem and social status.

Physical and occupational therapy

Physical therapy and occupational therapy are useful to improve movement quality and range of motion. Range of motion and strengthening exercises as well as neurofacilitation methods are part of treatment. Activities involving the use of both hands improve function. Provide adequate sensory stimulation to develop better hand control [E].

Inhibiting the normal extremity by bracing or casting and forcing the plegic one to work may be useful in the young child during the period of the development of hand-eye coordination.

Bracing

The effects of bracing are unclear. Night splints in functional position may promote lengthening of muscle-tendon units and prevent deformity. However most children sleep with a completely relaxed arm and extended hand which make night splints seem useless [F]. Neoprene thumb splints to keep the thumb out of the palm or thermoplastic wrist extension splints are commonly used during the day [G].

Local anesthetic and botulinum toxin blocks

Local anesthetic blocks are used to determine the presence of a contracture and to assess power in the antagonist muscles. Block the median nerve at the elbow to relax the flexor muscles in the forearm. Spastic muscles will relax completely after the median nerve block. If the wrist or the fingers remain flexed after the local anesthetic injection, this indicates a fixed contracture and will benefit only from surgery.

Check for active muscle contraction in the antagonist muscles. The presence of voluntary wrist and finger extension after the block indicates better functional prognosis after botulinum toxin injections or surgery to relieve flexor spasticity.

Dynamic contracture caused by spasticity responds well to botulinum toxin injections. This method is particularly valuable in the young child from age 2 to 6 years because relief of spasticity allows him to use the hand better. This may permanently improve hand function, sensation and hand-eye coordination. The dose is 1-2 units per kilogram of body weight per muscle. EMG or electrical stimulation guide is beneficial to target the spastic muscles, but this is a painful technique and requires conscious sedation or general anesthesia in most children except the very bright and courageous. Because botulinum toxin effects are temporary, consider surgical intervention in the older child for definitive treatment.

A Upper extremity problems

Lack of voluntary control
Poor hand-eye coordination
Sensory loss
Astereognosis
Spasticity
Dystonia
Weakness
Contractures
Joint instability

B Common deformities of the upper extremity

Shoulder	Internal rotation, adduction
Elbow	Flexion
Forearm	Pronation
Wrist	Flexion
Fingers	Flexion, ulnar deviation, swan neck
Thumb	Adduction, flexion

Spasticity and loss of selective motor control prevent positioning the upper extremity and manipulating objects with the hand.

Evaluate the hand using toys and simple every day tools. Determine the missing function and work towards mastering that.

F Resting splints to prevent deformity

Resting hand splint	Wrist: 30° extension, Metacarpophalangeal joints: 60° flexion Interphalangeal joints: neutral position Thumb: opposition
Ball abduction splint	Thumb abduction & opposition

G Splints to improve function

Wrist cock-up splint	Wrist: 30° extension Thumb: abduction Finger movement free
Soft thumb loop splint	Thumb: out of the palm
Opponens splint	Thumb: abduction & opposition Wrist: 30° extension

Surgery

Upper extremity surgery can improve hand function in a few selected cases [A]. The ideal surgical candidate must be a motivated, intelligent child who has good sensation in the hand and uses the extremity. Those children with satisfactory hand-eye coordination can benefit from surgery even when hand sensation is poor.

The surgeon must be careful in patient selection because some children develop adaptive mechanisms to compensate for lost hand movements as they grow. Functional loss occurs after surgery in such patients because surgery prevents the adaptive movements they developed over the years.

Consider surgery between 6- 12 years of age when the child will cooperate with postoperative rehabilitation. Set goals that fit with the expectations of the child and the parents.

The shoulder Adduction - internal rotation contracture is the most common problem. Provide a program of stretching exercises. Consider surgical lengthening of the muscles if the deformity is severe.

The elbow Flexion contractures of more than 45 are functionally disabling. Try botulinum toxin injection to elbow flexors and stretching exercises in dynamic deformities and even for cosmetic reasons. Consider surgery for elbow only if the hand is functional, if there is skin breakdown at the elbow or if hygiene in the antecubital fossa is poor. Deformities greater than 60° require surgical lengthening of the biceps tendon, be aware of the fact that this procedure worsens the forearm pronation deformity. Maximum range of motion is gained 3 months postoperatively.

Forearm The main problem is a pronation contracture because of spasticity in the pronator teres and pronator quadratus muscles [B]. Activities that require supination like grasping a walker or a cane, balancing objects in the palm, washing the face are impossible. Severe pronation causes radial head dislocation but it is generally painless and does not cause functional problems.

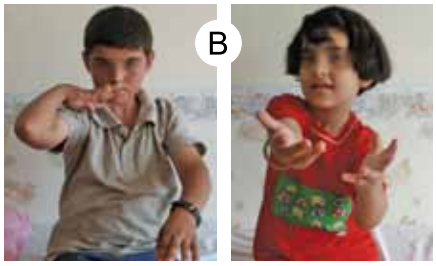
Consider pronator teres transfer to the supinator if the child can voluntarily pronate the forearm. Pronator release gives satisfactory results if the child has active supination. Long-standing pronation contracture of the forearm leads to relative shortening of the biceps aponeurosis. Release this structure to allow the biceps to be a more effective supinator [C].

Wrist The wrist usually is held in a position of flexion and ulnar deviation because of flexor carpi radialis and flexor carpi ulnaris spasticity [D, E]. The digital flexors also contribute to wrist flexion. Finger flexors are inefficient and the grasp is weak when the wrist is flexed [F]. Grasping is essential for function. Correct flexion contractures of wrist and fingers and adduction of thumb if they interfere with grasp. Macerations and mycotic infections are common in severe flexion contractures of the hand. Surgery becomes necessary for hygienic purposes.

Options for surgery [G] include wrist flexor lengthening, flexor origin slide, tendon transfer to improve wrist extension, proximal row carpectomy, and wrist fusion with or without carpal shortening [H]. Avoid wrist arthrodesis because the patient loses the tenodesis effect of wrist extension that results in finger flexion and facilitates grasp and release. Consider wrist arthrodesis only to relieve the pain and improve the cosmesis of the hand when there is no or limited hand function.

Wrist and digital flexor muscles can be selectively lengthened distally. Do not release or transfer both flexor carpi ulnaris and radialis as this eliminates active wrist flexion.

A	Before surgery consider
	Voluntary hand use
	Sensation
	Intelligence
	Athetosis



Limitation of forearm supination is a common problem of the hemiplegic upper extremity. It is also one of the most functionally disabling deformities.

C	Surgery for pronation contracture
	Release of lasertus fibrosis
	Release pronator teres insertion
	Pronator teres rerouting
	Flexor-pronator slide
	Pronator quadratus recession

Wrist flexion in hemiplegia may be a combination of spasticity and dystonia.

Wrist flexion impairs the ability to grasp objects and limits the use of the hand.

Treatment of wrist flexion deformity	G
Active finger extension at 20° wrist flexion: No need for surgery	
Active finger extension with the wrist over 20° flexion: Flexor releases, augmentation of wrist extensors or flexor carpi ulnaris release.	
No active finger flexion: Finger extensors must be augmented with flexor carpi ulnaris.	

Indications for wrist arthrodesis	H
No wrist control, strong finger flexion and extension	
Severe wrist flexion deformity and weak hand and wrist muscles	
Athetosis or dystonia, when finger function improves by wrist immobilization	

Consider tendon transfers to augment wrist extension when necessary. Transfer the flexor carpi ulnaris to extensor digitorum communis when both finger and wrist extension are weak. This transfer improves wrist extension and does not impair finger extension and release.

Fingers Finger flexion deformity is a result of spasticity and contracture in the flexor digitorum superficialis and profundus muscles [A]. It becomes more obvious when the wrist and metacarpophalangeal joints are held in neutral position.

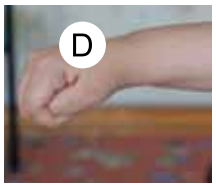
Consider surgical intervention when flexion deformity is severe [B]. The flexor-pronator origin release effectively lengthens the flexor digitorum superficialis, pronator teres and flexor carpi radialis. Correct finger flexion deformity by direct Z-lengthening of involved tendons. If there is spasticity of intrinsic hand muscles, releasing the finger flexors will increase the deformity. Excessive lengthening weakens flexor power, impairs grasp, and can produce swan neck deformities. In this case, transfer the flexor digitorum superficialis tendon to augment wrist, finger or thumb extension instead of lengthening.

Swan-neck deformity [C] is hyperextension deformity of the proximal interphalangeal joints caused by over-activity of the intrinsic muscles. This deformity increases with the pull of the extensor digitorum communis when the wrist is in flexion. Consider surgical intervention if there is severe hyperextension, or when the proximal interphalangeal joints lock in extension.

The thumb The thumb-in-palm deformity [D] is characterized by metacarpal flexion and adduction, metacarpophalangeal joint flexion or hyperextension and usually interphalangeal joint flexion [E]. The causes are spasticity and contracture of the adductor pollicis, first dorsal interosseous, flexor pollicis brevis, and flexor pollicis longus [F]. The extensor pollicis longus, extensor pollicis brevis, and/or abductor pollicis longus are often weak or ineffective. Thumb-in-palm deformity impairs the ability of the hand to accept, grasp, and release objects. The goals of surgery [G] are to release the spastic muscles to position the thumb, to create a balance in the muscles around the thumb, and to provide articular stability for grasp and pinch.

Procedures for finger flexion deformity B
Flexor-pronator origin release
Specific lengthening of musculotendinous units (Fractional lengthening or z-lengthening)
Sublimis to profundus tendon transfer
Finger flexor transfer for Wrist, finger, or thumb extension

Swan-neck deformity is generally not functionally disabling.



Thumb-in-palm deformity in a teenager.

The thumb-in-palm deformity E
Simple metacarpal adduction
Metacarpal adduction & metacarpophalangeal joint flexion
Metacarpal adduction with hyperextension instability of the metacarpophalangeal joint
Metacarpal adduction, metacarpophalangeal & interphalangeal joint flexion

Causes of thumb-in-palm deformity F
Contracture and spasticity of adductor pollicis, flexor pollicis brevis, flexor pollicis longus and first dorsal interosseous
Contractures of abductor pollicis longus, extensor pollicis brevis & extensor pollicis longus
Hypermobility of thumb metacarpophalangeal joint

Surgical procedures for thumb in palm deformity

G *Procedure*	*Location*	*Reason*
Tendinous insertion Muscular origin Fractional Release or lengthening	Adductor pollicis First dorsal interosseous Flexor pollicis brevis Flexor pollicis longus	Release contracture
Rerouting	Extensor pollicis longus	Augment active thumb abduction & extension
Arthrodesis Capsulodesis	Metacarpophalangeal joint	Stabilization
Four-flap Z-plasty deepening	Skin contracture	Between thumb & index fingers

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Diplegia

Diplegia is defined as gross motor involvement of the lower and fine motor involvement of the upper extremities [A]. Diplegia constitutes 50% of the spastic CP population. Diplegic children have normal mental function and can communicate without difficulty. Their oromotor and gastrointestinal functions are normal. They often have visual perceptual deficits and strabismus. There is a tendency to fall backwards because of poorly developed balance reactions.

The main problem in spastic diplegia is walking difficulty [B]. Balance disturbance, muscle weakness, spasticity and deformities result in abnormal gait patterns typical for diplegic children. Abnormal gait increases energy consumption causing fatigue. Most diplegic children start cruising at two years of age and walk by age four. Neuromotor function improves until age seven. Children who cannot walk by then in spite of appropriate treatment usually become limited walkers.

Among all types of CP diplegic children benefit most from treatment procedures. Unlike hemiplegic children they cannot fulfill their potential if left untreated. With treatment they may become productive members of the society. Every effort is worth spending when treating a diplegic child [C].

Physiotherapy and occupational therapy

Positioning, strengthening and stretching exercises preserve joint range of motion, increase strength and help improve gait [D]. Combine physiotherapy with bracing, walking aids and antispastic treatments to facilitate independent walking. The risk of contracture formation increases between ages 4-6 and during the prepubertal growth spurt period when the rapid increase in bone growth is not accompanied by a similar growth in muscle lengths. Relative muscle shortening causes contractures during this period. Biarticular muscles such as psoas, rectus femoris, hamstrings and gastrocnemius are more vulnerable. Intensive physiotherapy is then necessary to prevent contractures.

Diplegic children should receive physiotherapy until they are preschoolers. Boring exercises should be combined with play activities particularly in toddlers and in noncompliant children. Provide antispastic medications above age two if spasticity interferes with mobility and sleep. Time all orthopaedic interventions in the preschooler so that they do not interfere with the child's education. Sports activities and play with peers are essential during school years. Swimming and horseback riding are beneficial for the poorly developed balance reactions of the diplegic. These activities restore a sense of well-being and self-confidence in the child.

Provide occupational therapy to improve hand function if there are obvious coordination problems.

Botulinum toxin

Botulinum toxin is useful to relieve spasticity of the lower extremities of the young diplegic child. Consider injecting when spasticity becomes an obstacle to mobility and causes contractures [E]. Many muscles need injecting, do not exceed a total dose of 400 units in a single session. When the necessary dose exceeds 400 units, use phenol motor point block to the proximal muscles and botulinum toxin to the distal muscles. It is better to perform multiple muscle injections under general anesthesia or conscious sedation. Use simple local anesthetic creams beforehand for single muscle injections.

Deficient balance reactions and lower extremity spasticity are the main reasons of walking difficulty in diplegic children.

Musculoskeletal problems in diplegia B

Hip	Flexion, internal rotation and adduction
Knee	Flexion or occasionally extension
Ankle	Equinus, valgus (rarely varus)

Treatment in diplegia C

Physiotherapy	Increase strength Decrease spasticity Prevent contractures Improve gait
Occupational therapy	Improve hand function
Bracing	Solid or hinged AFOs or GRAFOs
Botulinum toxin	Decrease spasticity Hip: flexor/adductor Knee: flexor/extensor Ankle: plantar flexor/peroneal muscles
Orthopaedic surgery	Correct deformities

Stretching and strengthening exercises are fundamental components of physiotherapy in diplegia.

E **Botulinum toxin A injections in diplegia**

	Jump	*Crouch*	*Scissoring*	*Stiff knee*
Psoas	1 - 2 u	1 - 2 u	1 - 2 u	
Adductors	3 - 6 u		3 - 6 u	
Rectus femoris	3 - 6 u			3 - 6 u *
Medial hamstrings	3 - 6 u	3 - 6 u	3 - 6 u	
Lateral hamstrings	3 - 6 u	3 - 6 u		
Gastrocnemius	3 - 6 u			

The given doses are per kg of body weight. Total dose should not exceed 12 u/kg of body weight or 400 U.
*Inject all heads of the quadriceps muscle in stiff knee gait.

Casting after botulinum toxin injections is thought to enhance and prolong the effect. Continue with physiotherapy and bracing. The toxin has a temporary effect, yet it is an important tool to relieve spasticity in the young child when it is too early for orthopaedic surgery. Older children benefit from a combined use of botulinum toxin with surgery. Inject muscles without contractures and surgically lengthen those with contractures. This combined approach with surgery decreases the extent of surgery and enables a faster return of function in the postoperative period.

Bracing

Most diplegic children need variations of the AFO [A]. AFOs provide a stable base for standing and maintain good joint alignment during walking. Prescribe solid, hinged AFOs or GRAFOs depending on the gait pathology. Resting and night KAFOs are used to prevent knee and ankle contractures [B]. The child with severe spasticity cannot tolerate these, wakes up often and cries a lot. Do not use night splints if there is severe spasticity or contracture, relieve spasticity first.

Other measures

A small group of mildly involved diplegic children may benefit from selective dorsal rhizotomy. The ideal candidate for SDR is the independent ambulator between the ages of 3-10 with pure spasticity, good balance, no deformities, and a strong family support. The procedure is technically complex, and there is a need for long intensive physiotherapy afterwards. There may be increases in the hip and spinal pathology after the procedure. The long-term effects of SDR are still controversial though it has a place in treating spasticity in a very selective group of diplegic children.

Use of intrathecal baclofen pumps are becoming more common in ambulatory diplegic children. The complication rate and the expense limit their use.

Orthopaedic surgery

Most deformities of diplegics can be prevented or corrected with appropriate surgery. Therefore the most successful outcomes are seen in diplegic children. Delay surgery until the child is able to cruise holding unto furniture or walk holding hands. Provide intensive physiotherapy and botulinum toxin injections to lengthen the spastic muscles and prevent contractures during this period. The ideal age of operation is between 5-7 years. Early surgery is necessary in cases with hip instability, knee flexion contracture because of spastic hamstrings and contracture of gastrocnemius-soleus unresponsive to physiotherapy, botulinum toxin or serial casting.

Define clearly all of the musculoskeletal problems of the lower extremities prior to surgery and address them in a single setting in order to obtain a successful result. Multiple operations for each separate deformity add to the burden of the child and the family, lengthen the treatment period and cause multiple hospitalizations [C].

Multilevel surgery

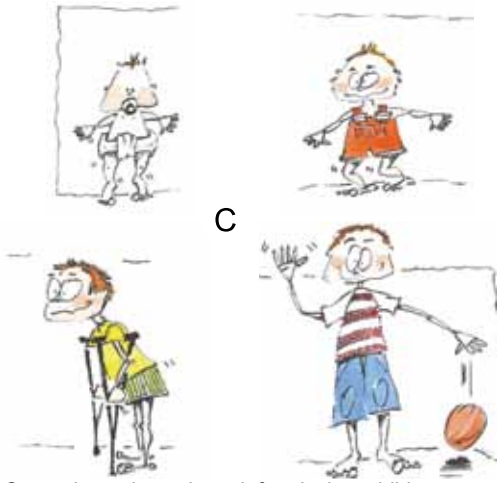
Multilevel surgery is performing multiple surgical interventions at a single session. This concept evolved when physicians realized that doing one operation at a time did not address the complex gait pathologies of CP [D]. Perform all surgery directed at the hip, knee and ankle such as hip adductor releases, hamstring and gastrocnemius lengthenings or rectus transfers simultaneously during a single session to correct jump, crouch, stiff knee or scissoring gait. Add bony procedures for deformities such as hip subluxation, femoral anteversion, external and internal tibial torsion and severe pes valgus. Prescribe intensive physiotherapy to strengthen the muscles, prevent contractures and increase function after multilevel surgery.

All children do not need multilevel surgery. Some have mild problems and require lengthening of one or two muscles only. Tailorize treatment according to the child's needs.

Type of brace	Indication
Hinged AFOs	Jump gait & 10° passive ankle dorsiflexion
Solid AFOs	Jump gait & no passive ankle dorsiflexion Crouch gait Severe pes equinovalgus
GRAFOs	Crouch gait
SMOs	Mild valgus-varus deformity & good ankle control

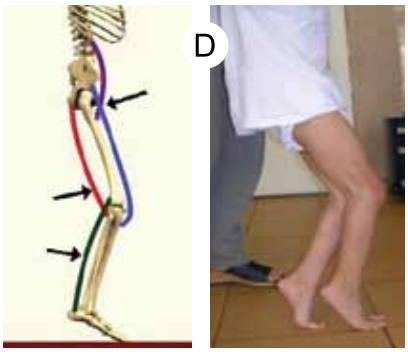
A

Use resting splints only after you reduce spasticity and make sure that the child sleeps well without waking up.



Correcting only equinus deformity in a child causes crouch gait with increased hip and knee flexion. A second operation to lengthen the knee flexors without addressing the hip flexors causes the child to bend at the hips because of the spastic iliopsoas. Erect posture is possible only after the third operation to lengthen the hip flexors. Lengthen all the flexors of the lower limb in a single surgical session to reduce hospitalizations.

Illustration by Idil Çilingiroğlu



Multilevel surgery is not the universal solution for every diplegic child. Some children need hamstring or gastrocnemius lengthening only. Plan surgery as the child's needs dictate.

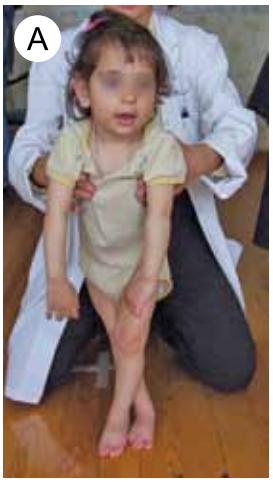
Musculoskeletal problems and their treatment

Muscle imbalance, spasticity and deformities at the hips, knees and ankles contribute to the specific posture and gait patterns typical for diplegic CP.

Scissoring

Scissoring is a frontal plane pathology also called crossing over. It occurs as a result of hip adductor and/or medial hamstring spasticity [A]. Persistent femoral anteversion is another important cause of scissoring. The child walks with legs crossing one another. [B] The hip is in flexion, adduction and internal rotation. The knees are turned inward. Scissoring gait may accompany sagittal plane pathologies such as jump or crouch knee gait.

Give stretching exercises to the hip adductors and medial hamstrings. Advise night splints for keeping the hips in abduction in the young child. W-sitting may increase adduction and internal rotation. It is presumed to reinforce femoral anteversion. However, if W-sitting is the only way the child can maintain sitting balance, do not prevent it. Encourage tailor- sitting or using an abduction wedge. Botulinum toxin injections in a dose of 50-75 units per muscle to the adductors and medial hamstrings temporarily increase range of motion. Adductor and psoas spasticity may result in hip subluxation. Lengthening tight hip adductors and medial hamstrings becomes necessary. Femoral derotation osteotomies are necessary if scissoring is caused by femoral anteversion.



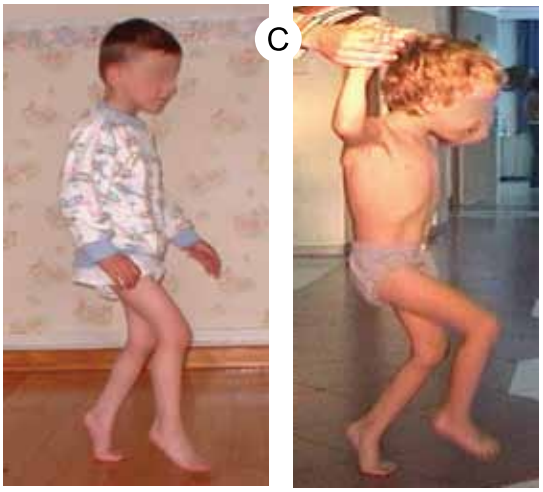
This type of scissoring is typical in the young diplegic child who is just beginning to walk.

Scissoring in the older child is because of persistent femoral anteversion, medial hamstring and adductor spasticity.

Jump gait

Jump gait is the most common sagittal plane pathology in young diplegic children. Almost all diplegic children begin walking with a jump knee gait pattern. Jump gait is defined as excessive hip flexion, knee flexion and equinus in stance [C]. The cause is lower extremity flexor muscle spasticity. The child walks with hips and knees in flexion and ankles in plantar flexion looking like an athlete getting ready to jump.

Early treatment consists of multilevel botulinum toxin injections to the hip, knee and ankle flexors in addition to aggressive physiotherapy and AFOs. Strengthen the weak lower extremity muscles (gluteus maximus, quadriceps and tibialis anterior) and stretch the spastic muscles. Most children with jump gait require surgery around the age of 5-6 to release tight hip flexors and lengthen knee and ankle flexors. Perform all operations at a single session. Combine with adductor releases at the hip if necessary.

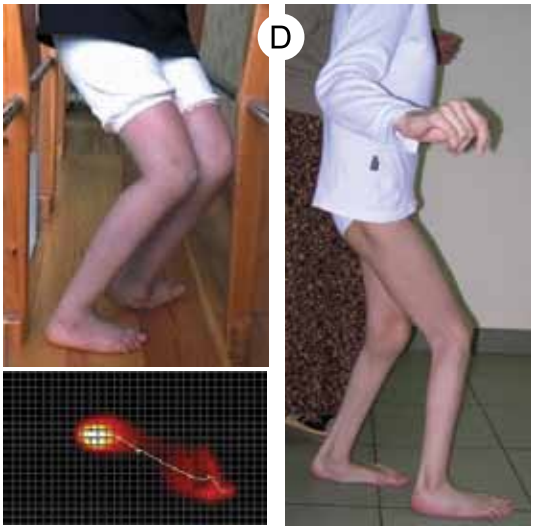


Younger diplegic children show a jump gait pattern with hips, knees and ankles in flexion when they first start walking. They need to hold hands or use a walker, rarely they can balance themselves.

Crouch gait

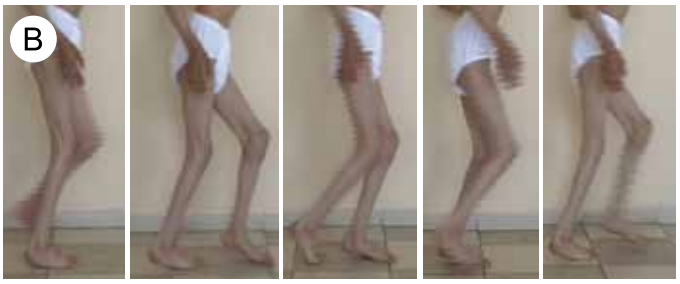
Crouch gait is the second most common sagittal plane pathology and it occurs in the older diplegic [D]. It is defined as excessive knee flexion throughout the stance phase with dorsiflexion of the ankle joint. Pes valgus generally accompanies crouch. Common causes of crouch gait are short or spastic hamstrings, hip flexor tightness and excessive ankle dorsiflexion. Excessive ankle dorsiflexion may result from isolated triceps surae lengthening without addressing the spastic hamstrings. Hamstring tightness causes crouch and a short step length when walking. When sitting, tight hamstrings pull the ischial tuberosities and tilt pelvis posteriorly causing kyphosis and sacral sitting.

Treatment of crouch gait is difficult. Nonsurgical treatment methods are physical therapy to stretch the hamstrings and strengthen the quadriceps and triceps muscles. A GRAFO is useful to bring the ground reaction force in front of the knee and create an extensor moment.



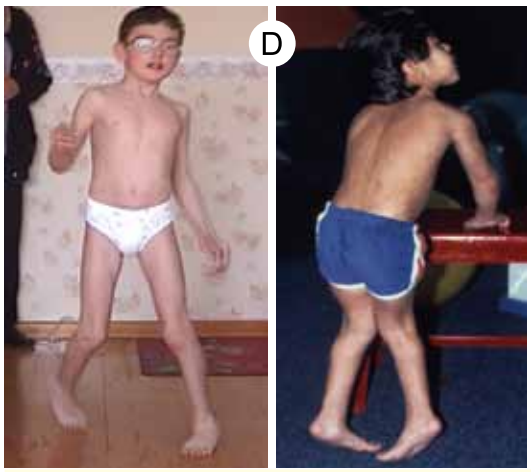
Crouch gait, common in the older diplegic child is characterized by increased knee and hip flexion with ankle dorsiflexion. Pedobarography shows the disturbed load distribution: the heel carries most of the body weight.

Knee flexion is the most common knee deformity in the diplegic child. It occurs in combination with hip flexion and ankle equinus.



Stiff knee gait is characterized by decreased knee range of motion during walking.

Genu recurvatum is usually seen as a secondary problem because of mild pes equinus.



Femoral anteversion leads to intoeing and pes equinus.

Miserable malalignment syndrome consists of femoral anteversion and external tibial torsion forcing the feet into valgus.

Lengthen the hamstrings in children who have hamstring shortening and/or knee flexion contractures. After surgery, strengthen the gluteus maximus, quadriceps and triceps muscles by intensive physiotherapy. Use GRAFOs to prevent excessive knee flexion postoperatively.

Hamstring contractures cause knee flexion deformity [A]. Supracondylar extension osteotomy may be necessary in severe cases.

Stiff knee

This is a sagittal plane pathology characterized by limited range of motion in the knee joint, especially a lack of flexion in swing [B]. It occurs because of spasticity of rectus femoris muscle or unopposed rectus femoris function after hamstring lengthening. Compensatory movements of hip external rotation and circumduction are observed. The patient experiences difficulty going up steps. Step length is shortened, foot clearance is poor, shoes wear out rapidly.

Conservative treatment of stiff knee gait consists of stretching the rectus femoris. Botulinum toxin injections or motor point blocks with phenol to the rectus femoris can temporarily decrease spasticity and allow knee flexion. Transfer of the rectus femoris tendon posteriorly to the gracilis or semitendinosus can improve knee flexion.

Genu recurvatum

Genu recurvatum occurs in the stance phase of walking [C] and is generally associated with mild equinus caused by triceps surae spasticity, excessive spasticity in the quadriceps, and weakness of the hamstring muscles or contracture of the hip flexors.

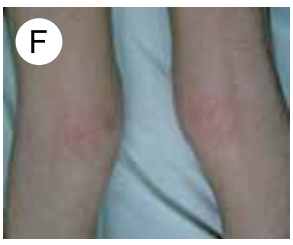
Botulinum toxin injections to the spastic gastrocnemius and rectus femoris muscles are useful in young children. AFOs set in 5 degree dorsiflexion prevent genu recurvatum. Transferring the spastic rectus femoris to the medial hamstring and lengthening the gastrocnemius muscle are surgical options.

Torsional deformities

Femoral anteversion is naturally increased in all babies and regresses as the child grows. Persistent femoral anteversion causes scissoring and intoeing gait [D]. Adductor and flexor tightness also contribute to scissoring caused by increased femoral internal rotation. The knee and ankle joints do not function on the plane of movement and walking difficulty is increased.

There is no conservative treatment for torsional deformities. Perform proximal or distal femoral derotation osteotomies to correct this problem.

Compensatory tibial external torsion is often secondary to femoral anteversion and causes pes valgus in many children. This 'malignant malalignment syndrome' [E, F] requires external rotation osteotomy of the femur along with internal rotation osteotomy of the tibia.



Skin irritation at the medial side of the femoral condyles because legs rub against each other.

Hip

The risk of hip instability is less in diplegics than in the total body involved children. All diplegic children should still have baseline radiographs. Adductor stretching, positioning, and botulinum toxin injections decrease spasticity to a certain extent. Surgery is necessary in children with hips at risk.

Pes valgus

Pes valgus is characterized by abnormal eversion of the heel [A], convexity of the medial border of the foot [B] and prominence of the head of the talus [C]. It occurs because of spasticity of the peroneals, extensor digitorum communis and triceps surae. External tibial torsion creates a valgus stress at the ankle and contributes to pes valgus [D].

The natural history of certain mild developmental problems of the lower extremities such as pes planovalgus and genu recurvatum is benign. These disorders are seen in able bodied children as well and disappear spontaneously around 7-8 years of age as ligaments get tighter.

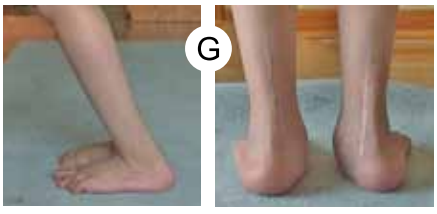
Severe pes valgus deformity causes callosities on the medial side of the foot, midfoot abduction and hallux valgus. Exercises and casting are not effective. Orthopaedic shoes or shoe inserts cannot correct the deformity. UCBL, SMO or rigid AFOs with UCBL soles (footplates) provide a stable base for standing. Surgical options are limited. Lengthen the gastrocnemius if the pes valgus is because of gastrocnemius tightness. Use AFOs postoperatively.

Tendon transfers do not correct the muscle imbalance in pes valgus. Combine lengthening the peroneus brevis muscle with bone surgery in young children. Dennyson-Fullford subtalar arthrodesis or calcaneal neck lengthening (Evans procedure) preserve hindfoot mobility without disturbing the growth potential. Delay bone surgery until preadolescence except for cases with severe deformity and rapid progression of hallux valgus. Triple arthrodesis in adults and adolescents is a last resort.

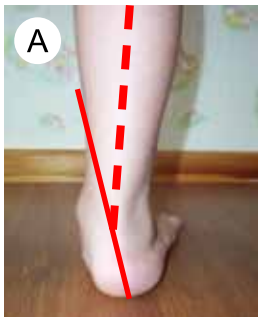
Hallux valgus

Hallux valgus occurs secondary to pes valgus or pes equinovalgus in ambulatory children [E]. Correct equinovalgus deformity first, hallux valgus deformity improves after this. Spasticity of the adductor hallucis muscle causes hallux valgus in plantigrade feet [F]. In this case, release the spastic muscle. Comfortable shoes with a wide toe box are useful for mild deformities. Perform metatarsal osteotomies or metatarsophalangeal arthrodesis in severe cases.

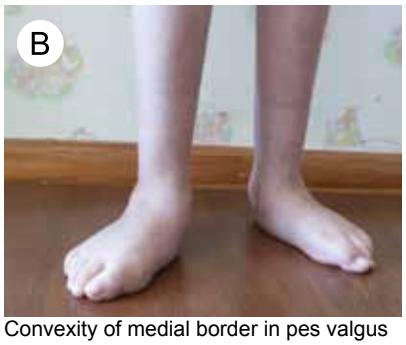
The foot and ankle problems of the child with CP must be evaluated as a whole, not as separate deformities. A problem in one joint leads to problems in all the others [G,H]. Do not intervene unless you are certain of the effects of your intervention on all the other joints of the extremities.



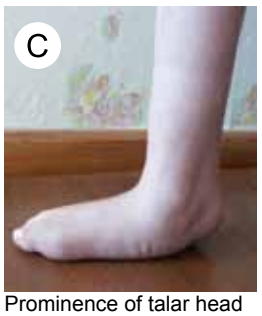
This child has hallux valgus secondary to crouch gait. An Achilles tendon lengthening caused pes calcaneus that led to crouch when combined with untreated hamstring spasticity. Treatment plan should include hamstring lengthening and bilateral GRAFOs.



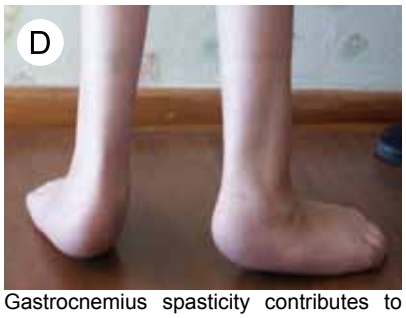
Hindfoot valgus



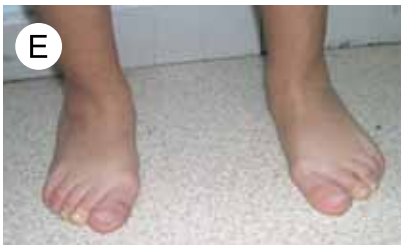
Convexity of medial border in pes valgus



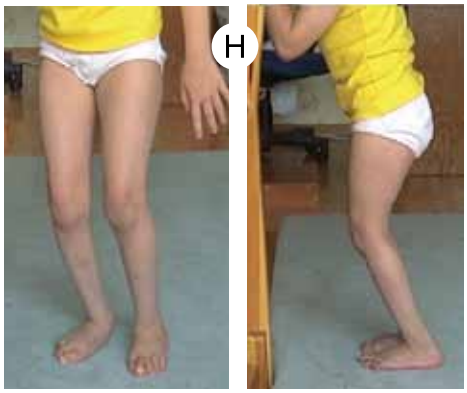
Prominence of talar head



Gastrocnemius spasticity contributes to pes valgus.



Hallux valgus in the younger and older child



Postoperative care of the diplegic child		
	Muscle-tendon surgery	*Bone surgery*
Pain relief	Epidural or caudal analgesia, antispastic medication, NSAIDs, narcotics	
Immobilization	3 - 6 weeks in bivalved casts or splints, 6 weeks in cast for tendon transfers at the foot	3 weeks in cast, no need for cast in the older child after femoral derotation osteotomy
Physiotherapy	Intensive for 3 months, strengthening and range of motion exercises, switch gradually to swimming and sports	
Ambulation	2 - 4 days postoperatively	3-6 weeks

A

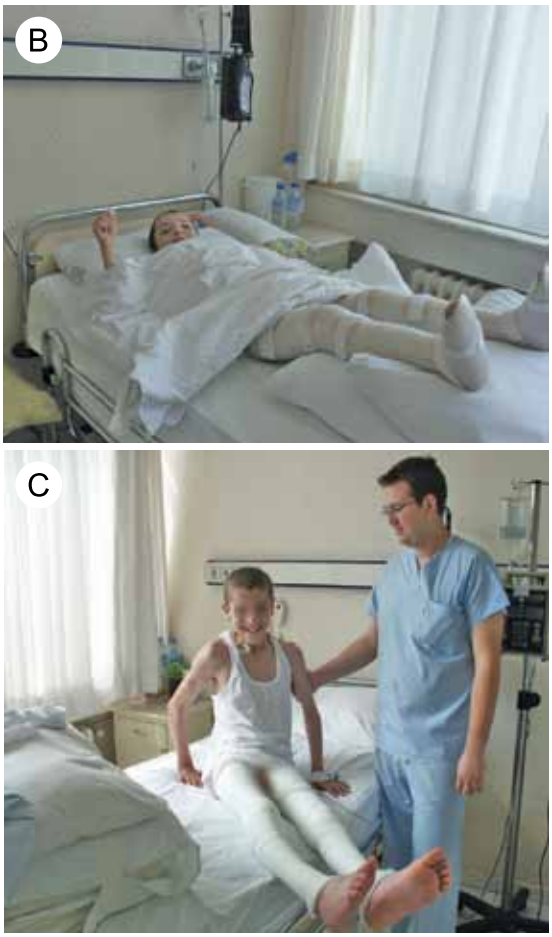
Postoperative care

Postoperative care of the diplegic child consists of pain and anxiety relief, antispastic medication, early mobilization, bracing and intensive physiotherapy [A]. Epidural analgesia is helpful in the early period after surgery [B]. Oral baclofen or diazepam decrease muscle spasms and pain. Use plastic KAFOs or combine knee immobilizers with AFOs to immobilise the lower extremity and allow ambulation on the second to third postoperative day after muscle tendon lengthenings [C]. The importance of strengthening the lower extremity muscles, especially those that have been lengthened cannot be overemphasized. Begin active exercises and sports after 6 weeks, as the child's general medical condition allows. Swimming, riding a bicycle or a tricycle, and playing ball are excellent options. Progress from parallel bars to a reverse walker with wheels and to forearm crutches or gait poles depending on the child's balance. Do not neglect strengthening and range of motion exercises in the first 3-6 months after surgery.

The beneficial effects of the surgical intervention become obvious in the first 6 months after surgery, the child continues to progress for up to one to two years postoperatively. Neglected cases have a longer recovery period.

Upper extremity

The upper extremity of the diplegic child is generally free from deformity. Severe cases have difficulty with fine motor control, they are slow and clumsy in activities of daily living, self-care and writing. These children benefit from occupational therapy to improve hand function.



Elevation of the lower extremities, patient controlled epidural analgesia and early mobilization allow a faster return to function.

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Quadriplegia

Quadriplegia is the involvement of neck, trunk and all four extremities. Quadriplegics have severe motor impairment and other signs and symptoms of CNS dysfunction such as cognitive impairments, seizures, speech and swallowing difficulties [A]. Some call this total body involvement because the trunk, neck and orofacial muscles are affected as well as the extremities.

Primitive reflexes persist, extrapyramidal signs such as dystonia and athetosis are common. Mental retardation, seizures, visual deficits, strabismus, bulbar dysfunction manifested by drooling, dysphagia, dysarthria and medical complications are frequent [B,C]. Gastroesophageal reflux causes feeding difficulty and can result in aspiration pneumonia [D]. Growth retardation is typical in severe cases. Many do not have bladder and bowel control. Cerebral dysfunction is more extensive and prognosis is worse. The spectrum of severity is variable, from having no sitting ability or head control to being able to walk independently. With proper treatment and education, children who have adequate mental function can use a wheelchair and communicate through a computer or other alternative aids. The majority of quadriplegics cannot be independent and need assistance in daily life. Only about 15% have the potential to walk and the rest are wheelchair bound. Most of them require lifelong all day care by the family. Most severely involved non-ambulatory quadriplegic children do not survive beyond adolescence. The survivors face the late complications related to hip instability and spinal deformity.

Spine and hip deformities such as hip instability, pelvic obliquity and scoliosis are very common and interfere with sitting balance [E]. Knee and ankle deformities seen in hemiplegic and diplegic children may also exist in quadriplegia. The incidence of lower extremity contractures increase with severity of the motor impairment.

Goals of treatment

Management strategy changes over time. Between ages 0-2 years, emphasize physiotherapy, infant stimulation, positioning and parent education. During ages 2-5 muscle tone becomes a problem, dyskinesias manifest themselves. Look for ways to decrease muscle tone. From 5 years onwards consider orthopaedic interventions. During the teen years provide better hygiene and seating for the nonambulator; prevent pain secondary to spasticity.

The main goal is to obtain and maintain sitting balance [F]. Good sitting in the upright position facilitates care, enables independence with a motorized chair and frees the hands for any limited use. The child can become partially independent in activities of daily living. Stable hips and a straight spine are necessary to sit independently in the wheelchair. Prevent deformity in the spine and hip, correct the existing deformities, try to preserve standing ability for transfers.

Total body involved spastic children generally cannot walk, often need seating supports, have spinal and hip deformities and many other medical problems which complicate the management.

Associated problems in quadriplegia (B)
Mental retardation
Seizures
Dysarthria-dysphasia
Incontinence
Hydrocephalus
Deafness
Visual impairment
Gastrointestinal disorder

Mental retardation, communication difficulty, drooling and dysphagia coexist in quadriplegia.

Gastrostomy is helpful in children with difficulty swallowing and severe gastroesophageal reflux.

E	Musculoskeletal problems in quadriplegia	
	Spine	Scoliosis Hyperkyphosis
	Hip	Subluxation Dislocation
	Knee	Flexion
	Ankle	Plantar flexion

Treatment in quadriplegia (F)	
Physiotherapy	Prevent hip subluxation Decrease deformity Preserve cardiovascular fitness
Occupational therapy	Provide assistive aids Adaptive equipment Increase independence in ADLs
Bracing	Spinal braces for better sitting Hip abduction brace for hip stability Resting splints for the knee & ankle
Seating aids	Proper positioning
Spasticity management	Oral medication Intrathecal baclofen pump Botulinum toxin
Orthopaedic surgery	Correct spine and hip problems

Physiotherapy and occupational therapy

Neurofacilitation techniques like Vojta or Bobath are used with the hope of stimulating the CNS towards normal development during infancy. Mobility issues arise as the baby becomes a child. Some children try to pull to stand whereas others move around hopping on their backs like a bunny or crawling backwards. There is a group of severely affected children who are not motivated to move or have no ability to move by themselves [A]. Encourage every child to stand in a suitable stander for short periods during the day regardless of the ambulation potential [B]. The child will be able to see the world vertically and have a feeling of what it's like to be standing on his feet. Standing may prevent contracture and improve cardiovascular, bowel and bladder function. It may increase bone mass and decrease fracture rate.

Less severely affected patients gradually learn to stand independently. The ability to stand independently for short periods and to take a few steps increases independence in daily living activities to a great extent. Some severely involved children who have motivation to move should use a wheeled mobility device [C]. They can learn transfers and wheelchair activities. Provide powered mobility devices to children from 2 years of age. Continue physiotherapy in the preschool and school period to prevent contractures, strengthen the upper extremity and improve cardiovascular capacity. Also provide occupational and speech therapy to improve hand function and communication to children who need support.

Bracing

The quadriplegic child spends almost his entire day in the wheelchair [D]. The wheelchair must be very comfortable. Do not use the wheelchair as a stretching device.

Night splints to prevent knee and ankle contractures are poorly tolerated by the child. Contoured seating aids increase sitting balance. Prefer powered wheelchairs because they conserve energy and are easier to use. Quadriplegic children with intact cognitive function can learn wheelchair skills.

Use plastic rigid KAFO's for therapeutic ambulation in parallel bars. Parapodiums and gait trainers are available to assist walking in mildly involved quadriplegic children.

Orthopaedic treatment

Hip instability and spinal deformity are the most important orthopaedic problems of the nonambulatory quadriplegic child. They do not respond to conservative measures and generally require orthopaedic surgery. Knee and ankle flexion deformities of the ambulatory quadriplegic child should be treated according to the same principles as in diplegia.

Scoliosis

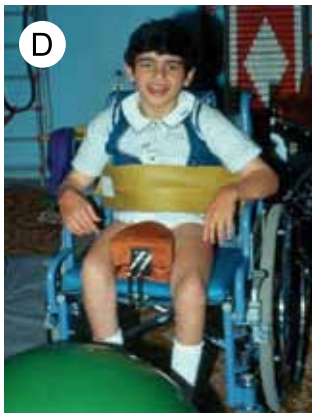
Scoliosis is the most common spinal deformity [E]. The incidence and severity varies directly with the severity of motor involvement. Quadriplegics are 10 - 15 times more prone to develop scoliosis than diplegics. Scoliosis causes difficulty with sitting and impairs breathing. Pressure sores and pain cause a further decline in the life quality of the individual.

Natural history Keep in mind that scoliosis in CP is different from idiopathic scoliosis [A on next page]. Scoliosis develops by age 5 to 6 in CP and is progressive. The deformity continues to progress after skeletal maturity, especially if the curve exceeds 40º. It cannot be controlled by orthotics and requires surgical treatment. Risk factors for curve progression are younger age, poor sitting balance, pelvic obliquity, hip dislocation and the presence of multiple curves [B on next page].

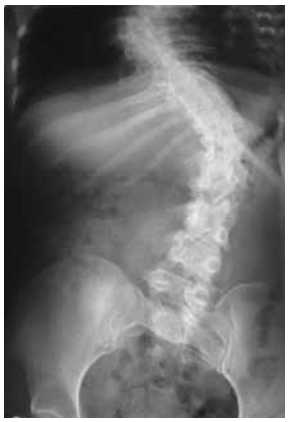
Some severely involved children do not have the motivation to sit by themselves and need external support in all positions.

Child supported in the near-vertical position in a stander develops a sense of verticality as a preparation for ambulation.

Associated problems such as visual impairments prevent mobility in the quadriplegic child.



Child sitting supported in the wheelchair. Ideally the wheelchair should become part of the child. Courtesy of G. Koloyan



Scoliosis interferes with sitting and also causes hip problems. It is the most common spinal pathology in quadriplegic children.

Conservative treatment The goal of treatment is to preserve the ability to sit erect and comfortably [C]. Good sitting improves the patients respiratory function, feeding, gastrointestinal function, hand use, mobility and communication. Do not operate on small curves that do not disturb sitting ability or large curves in severely involved patients. Provide a thoracolumbosacral brace (TLSO) in curves of 30º to 60º to slow curve progression and allow the spine to grow before surgical stabilization. TLSOs may improve sitting balance, particularly for those patients in whom surgery is not indicated and for those who still have significant spinal imbalance after surgical treatment.

A TLSO is the most effective and economical means of providing improved trunk support. Place a custom molded seating device inside the wheelchair for patients who cannot tolerate the TLSO. Simple wheelchair modifications may lessen progression, delay surgery to allow for spinal growth prior to fusion and enable proper sitting.

Surgical treatment Progressing scoliosis needs surgical stabilization [D]. Surgical correction of a high grade scoliosis in a total body involved child or young adult is difficult and may require anterior and posterior procedures [E]. Perform posterior spinal fusion with segmental instrumentation to achieve a balanced spine over a reasonably level pelvis. Perform segmental instrumentation with arthrodesis (fusion) of the spine to the pelvis to correct for pelvic obliquity [F,G]. Aim to achieve spinal balance in both the coronal and sagittal planes to maximize sitting balance. Extend the fusion to the upper thoracic region to minimize the risk of developing cephalad junctional kyphosis. Include the pelvis in the fusion if pelvic obliquity exceeds 10º from the intercrestal iliac line to the top of L5 or L4 when measured on a sitting anteroposterior radiograph. Perform fusion from the upper thoracic region (T1-T3) to L5 or to the pelvis. Pelvic obliquity continues to progress if not fused. Rarely a lesser degree curve can be treated without pelvic fusion [H].

Postoperative care There is no need for postoperative bracing. Have the patients sit in the upright position a few days after surgery. Be aware that these children are malnourished, prone to infection, and have difficulty communicating their needs and pain. Spasticity prevents appropriate positioning. Early postoperative mortality and morbidity is high. Preoperative nutritional status is important because malnourished patients have significantly higher infection rates and longer hospitalizations. Patients requiring both anterior and posterior fusions have fewer complications if both procedures are performed on the same day rather than at 1-to 2-week intervals. The surgeon's skill, speed, and stamina as well as patient blood loss and other factors determine the wisdom of same day anterior and posterior procedures in neuromuscular scoliosis.

Scoliosis in CP A

Develops by age 5 - 6
Progresses after skeletal maturity
Cannot be prevented by braces
Requires surgical treatment
Worsens quality of life
Shows poor prognosis

Risk factors for curve progression B

Younger age
Poor sitting balance
Pelvic obliquity
Hip dislocation
Presence of multiple curves

Requirements for comfortable balanced sitting & independent transfers C

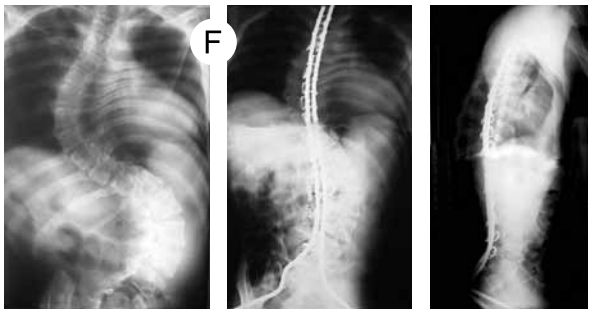
A straight spine and horizontal pelvis
Hip range of motion: 30º to 90º of flexion
Stable and painless hip
Knee range of motion: 20º to 90º of flexion
Plantigrade feet

Surgical indications for scoliosis D

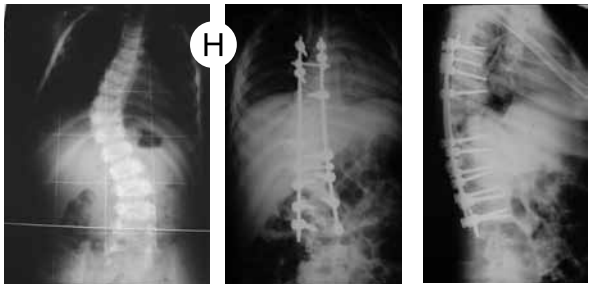
Curves > 50º
Fast curve progression
Pain
Deterioration of function

Treatment in different types of curves E

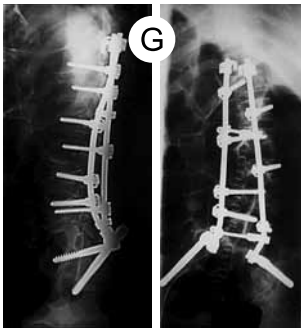
	Group I	*Group II*
Curve type	Double thoracic or thoracolumbar	Large lumbar or thoracolumbar
Pelvic obliquity	Little	Marked
Surgical procedure	Posterior fusion alone	Combined anterior & posterior fusion
Include pelvis in fusion	Nonambulatory patients only	All patients



Sublaminar wires attached to the laminae provide secure fixation. The rods can be extended to include pelvis in the fusion. Implant costs are minimum.



Use multiple pedicle screws for better correction and shorter fusion area if the dorsal skin coverage is adequate.



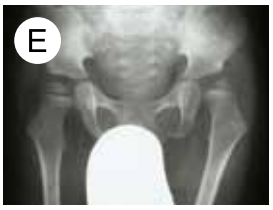
Another method of obtaining lumbopelvic fusion is attaching iliac screws to spinal rods.

Long C-shaped hyperkyphosis in the child with weak spinal extensor muscles

Hamstring spasticity causes posterior pelvic tilt and sacral sitting resulting in lumbar kyphosis.

Hip dislocation results in
Pain
Secondary scoliosis
Loss of sitting balance
Contralateral adduction deformity
Difficulty caring for the child dressing hygiene feeding

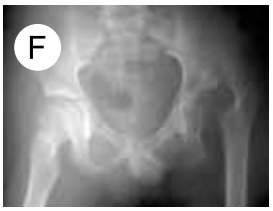
C



Hip subluxation is progressive unless treated.

D

Hip instability	
Hip at risk	Abduction <45° bilaterally or less abduction on one side
Hip subluxation	Femoral head migrates partially out of the acetabulum
Dislocation	All contact lost between femoral head & acetabulum



Hip dislocation causes difficulty with sitting and pain. The affected leg is shorter on examination.

Differences between developmental hip dysplasia & hip instability in CP		
	Developmental hip dysplasia	*Hip in CP*
At birth	Pathological	Normal
Dislocation	First months	After age 2
Etiology	Idiopathic	Secondary to CP
Pathophysiology	Progressive acetabular deficiency leading to dislocation	Spasticity, muscle imbalance, primitive reflexes & no weight bearing leading to progressive instability
Natural history	Moderate to poor	Poor to very poor
Treatment outcome	Good	Limited

G

H

Causes of instability
Muscle imbalance
Persistence of primitive reflexes
Absence of weight bearing

Hyperlordosis

Increased lordosis in the lumbar spine is usually secondary to hip flexion contractures and responds to correction of those contractures by appropriate means such as stretching or more often hip flexor release. Attempt spinal fusion and instrumentation to correct the deformity if it becomes rigid. Hyperlordosis can also be a compensatory deformity below a rigid thoracic hyperkyphosis, and it usually responds to correction of the primary problem.

Hyperkyphosis

Hyperkyphosis occurs in the young child with weak spinal extensor muscles [A]. There is a long, C-shaped forward posture of the entire spine. Correct this posture with proper seating, restraint straps on the wheelchair or a thoracolumbosacral orthosis providing support. A similar kyphosis occurs secondary to neglected hamstring contracture in the sitting patient. The hamstrings pull the pelvis and cause posterior pelvic tilt. The patient sits on his sacrum [B]. Lumbar lordosis decreases and thoracic kyphosis increases. Lengthen the hamstrings to correct this problem.

The hip

Hip dislocation affects hygiene, sitting, and gait of the total body involved child. It causes pain by early adulthood [C]. Secondary scoliosis and contralateral adduction deformity causing 'windswept hips' further worsen the situation. Dislocated hips are difficult to treat, emphasize early treatment to prevent progression of hip instability.

Classification Hip instability is classified as "hip at risk", hip subluxation and hip dislocation [D]. A "hip at risk" is defined as limitation of abduction to less than 45° bilaterally or markedly less abduction of one hip compared to the other. Hip subluxation is identified radiographically when the femoral head migrates partially out of the acetabulum [E]. Dislocation is present when all contact is lost between the femoral head and the acetabulum [F].

Pathogenesis and natural history The pathophysiology of hip instability is different from developmental hip dysplasia (DDH), the natural history is worse, outcome of salvage operations for the skeletally mature patients with a neglected hip are not always satisfactory. In contrast to DDH, the hips are normal at the first years of life. Progressive instability occurs later because of a combination of muscle imbalance, persistent primitive reflexes, faulty posture and absence of weight-bearing stimulation on bone [G].

The adductors and iliopsoas are spastic, causing adduction and flexion contractures. Hamstring spasticity contributes to muscle imbalance. Excessive muscle tone exerts a constant force on the developing hip, deforming both the femur and the acetabulum. Deformities include femoral anteversion (normal decrease in anteversion does not occur during early childhood, fetal anteversion persists) and coxa valga (increased neck shaft angle of the proximal femur). The combination of these abnormalities leads to acetabular insufficiency and hip instability [H]. The majority of the hips subluxate in the posterior-superior direction, because the adductors and flexors are stronger than abductors and extensors.

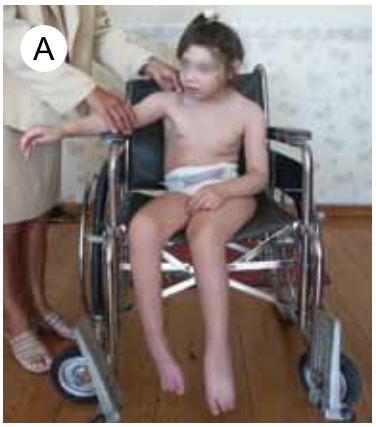
Hip subluxation usually begins between the ages of 2-6 years though dislocation can occur as early as 18 months of age. Most hips dislocate by 6 years of age if they are going to do so. Children with the most severe neurologic involvement have the

worst hips. The highest risk group is those who never achieve the ability to sit independently. The risk of hip instability is markedly less in diplegia and hemiplegia.

Clinical evaluation and follow up Perform a clinical examination of the hips and obtain radiographs in every child. Asymmetric sitting and a shorter leg are clues to underlying hip subluxation/dislocation [A,B]. Evaluate the hip abduction range both in flexion and extension [C,D]. Use the Thomas test to measure hip flexion contracture. Evaluate rotation in the prone position. Excessive femoral anteversion worsens the progression of hip instability. Hip instability is always progressive [E]. Monitor progression carefully. Test and record hip abduction. Repeat clinical and radiographic evaluation twice a year between the ages of 2 - 8. Baseline AP hip radiographs are obligatory in all diplegic and quadriplegic children. Measure the migration index (MI) on hip radiographs [F]. The upper limit of normal for the migration index is 20 % at age four. Computerized tomography with three-dimensional reconstruction is not essential but it shows deformities of the femoral head and the area of greatest acetabular deficiency (posterosuperior in most- but not all-cases). One can also measure femoral anteversion on computerized tomography.

Conservative treatment Prescribe physical therapy to all children to preserve hip motion and promote weight bearing. Physical therapy alone does not prevent hip subluxation. Use abduction splints or a pillow to keeps the knees apart. Botulinum toxin A can be injected in the adductors to temporarily decrease tone for 4 - 6 months.

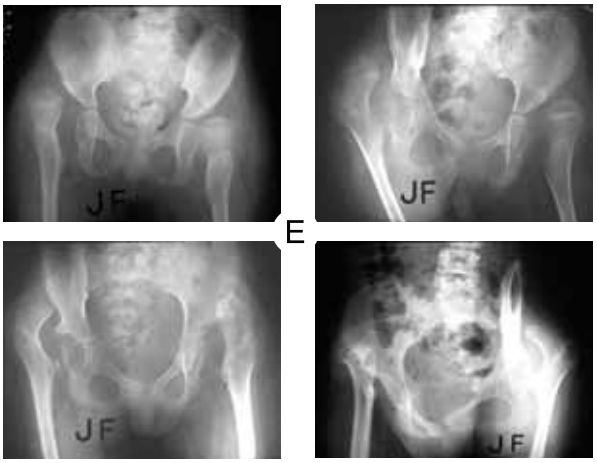
Adductor muscle lengthening Intervene early and release the hip adductor muscles to prevent the need for complicated hip reconstruction later. Adductor release is necessary if the migration index (MI) is greater than 20 % in children with scissoring or in any child with MI between 20 - 50 %. Consider adductor lengthening in children under age 4 even if MI is up to 75 % [G]. Strive to gain at least 60^{o} passive abduction on each side with the hip and knee flexed 90^{o} or at least 45^{o} abduction with the hip and knee extended. Dividing only the adductor longus is usually sufficient. Release the adductor brevis and gracilis muscles if necessary. Prefer open release to percutaneous techniques. Do the procedure bilaterally to balance the pelvis. Perform a fractional lengthening of the iliopsoas or a tenotomy if there is concomitant flexion contracture. Consider lengthening the rectus femoris muscle and the hamstring if popliteal angle is > 45^{o} and hamstring tightness contributes to hip instability. Use traction or an abduction pillow after adductor lengthening. Do not attempt obturator neurectomy. There are risks of overcorrection and hip abduction contracture.



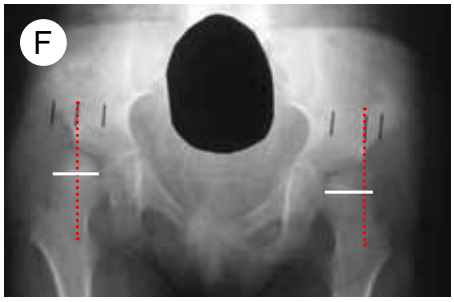
Hip subluxation disturbs sitting balance and leads to discomfort.

Leg length discrepancy is a sign of hip dislocation.

Examine hip abduction in flexion and extension. Obtain hip X-rays with 6 months intervals if there is persistent hip flexion or adduction tightness.



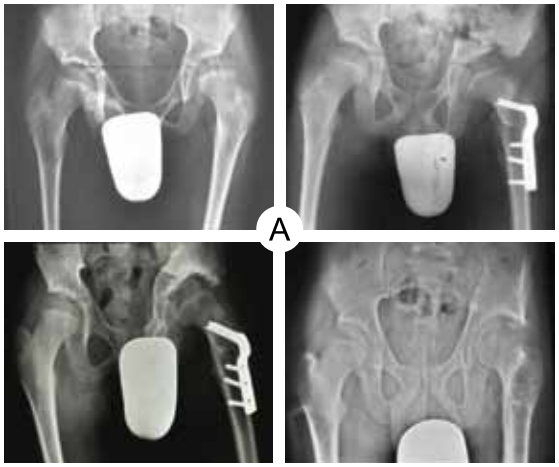
Neglected hip instability usually has a bad prognosis. The subluxed hips gradually dislocate, shortening gets worse, the high riding femoral heads disturb sitting and transfers. Intervene as early as is necessary to lengthen the spastic muscles. A minor operation saves the patient from extensive hip surgery later.



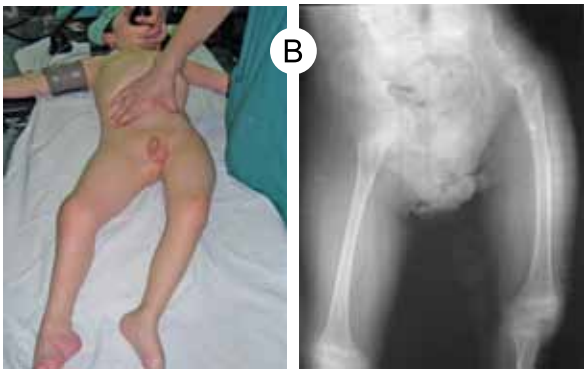
The Reimer's index: Draw a perpendicular line from the lateral acetabular margin. The percentage of the femoral head that lies lateral to this line is the migration index.

G

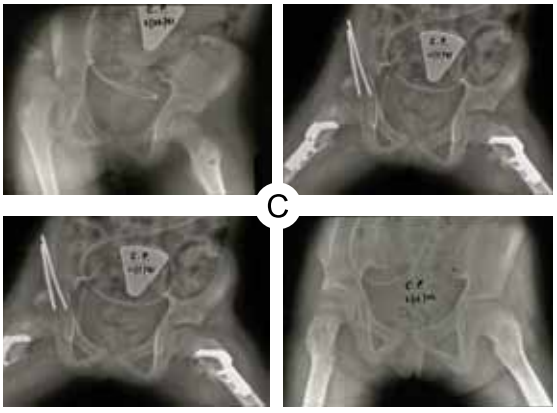
Treatment of the hip at risk	
Migration index	*Surgical procedure*
> 20 %	Follow-up
> 20 % + scissoring	Adductor +/- iliopsoas lengthening
20 - 50 %	
50 - 75 %, age < 4	
50 - 75 %, age < 4	Bony reconstruction
> 75 %	



Long term follow up of surgical treatment of a patient with hip subluxation. The combined femoral - iliac osteotomy and soft tissue releases have produced a stable and pain free hip joint.



The 'windswept hip' is the combination of hip dislocation and adduction deformity on one side and secondary abduction deformity on the contralateral hip.



The 'windswept hip' can only be treated by a series of major operations performed in the same session. The outcome can be excellent but the operation is traumatic for the child. Try to prevent hip instability from progressing to this advanced stage with simpler measures like early adductor releases.

The hip spica cast is prone to complications such as pressure sores. Careful follow-up is necessary.

Bone Surgery Perform a hip reconstruction when instability progresses after muscle lengthening, there is severe subluxation (MI > 75 %) or the hip is dislocated. Bony reconstruction is more reliable than adductor lengthening in children older than age four with an MI > 50% .

The age for hip reconstruction is 4 years and onwards. Older children have better bone stock for plate fixation. The upper age limit depends on the degree of the loss of sphericity of the femoral head. Hip reconstruction is successful before permanent advanced deformity of the femoral head occurs. Once the femoral head begins to flatten medially and laterally, loss of articular cartilage is likely and pain relief after reconstruction is not satisfactory.

Treatment of the subluxed hip The usual surgical procedure is a combination of femoral varus - derotation osteotomy, iliac osteotomy, capsuloplasty, adductor and iliopsoas lengthening [A]. Some of these are not necessary in certain children. Plan the procedures according to the needs of the child. Preoperative three dimensional CT scans may help surgical planning.

Treatment of the dislocated hip There are a large number of different techniques to reconstruct the severely subluxated or dislocated hip. The surgeon has to decide on the extent of surgery depending on the patients' pathology. In spastic hip disease, the acetabulum has a very limited ability to remodel once advanced dysplasia has developed. Because acetabular deficiency is posteriorly located in most cases variations of the Dega acetabuloplasty in combination with soft tissue lengthenings, femoral shortening, varus derotation osteotomy of the femur (VDRO) and capsuloplasty are preferred.

Windswept hip Treatment of the windswept hip [B] is a major task. The combined procedure is a femoral varus derotation osteotomy with shortening, iliac osteotomy and flexor adductor release on the dislocated and adducted side. This must be accompanied by a femoral osteotomy and soft tissue releases on the contralateral abducted side [C].

Salvage of the neglected dislocated or irreducible hip Painful hip subluxation or dislocation in the older child is difficult to treat, attempting to reduce the hip may be impossible. The salvage procedures for these children are resection arthroplasty, valgus osteotomy, arthrodesis and arthroplasty. Proximal femoral resection arthroplasty involves interpositioning of the muscles and capsule, is easier to perform and the aftercare is more comfortable both for the family and the surgeon. Valgus osteotomy is not universally accepted. Arthrodesis of the hip can provide a stable and painless hip but is a major procedure and often not well tolerated because of the long immobilization in a hip spica cast. Total hip replacement has been successful even in young children but should be done by someone who has experience in hip replacement as well as cerebral palsy.

In children who are able to stand for transfers and daily life activities or who are therapeutic ambulators, total hip arthroplasty provides a better outcome.

Postoperative care The patient is kept in a hip spica cast [D] for 4 - 8 weeks depending on the extent of surgery, bone quality, age and compliance.

The knee, ankle and the foot

There are some mildly involved quadriplegic children who have the potential to stand independently and take a few steps. Correct the knee and ankle deformities [A,B] in such children to enable efficient transfers and limited ambulation. Even limited ambulation can ease the caregiver's burden enormously, if a quadriplegic patient can stand to transfer, try to maintain this ability. Aim to obtain a comfortable posture in lying, sitting and in the standing frame. The knee should flex to 90° for sitting and extend to at least 20° for transfers. Severe knee flexion deformity causes skin sores behind the knee because of friction against the chair. Begin stretching and range of motion exercises early to prevent knee flexion deformity. Consider early hamstring lengthenings in children with deformity. Prescribe regular exercises, night splints and standing in the stander to protect the range of motion gained by surgical intervention. Distal femoral osteotomy is an option in children with knee flexion contractures and who have walking potential.

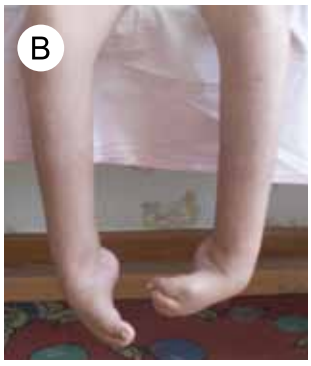
A plantigrade foot is necessary for standing during transfers and in the stander, placing the foot comfortably on the footrests in the wheelchair and wearing shoes. Stretching, range of motion exercises and orthotics may prevent deformity in the young child. Perform soft tissue procedures, corrective osteotomies or arthrodeses in the older child.

Upper extremity

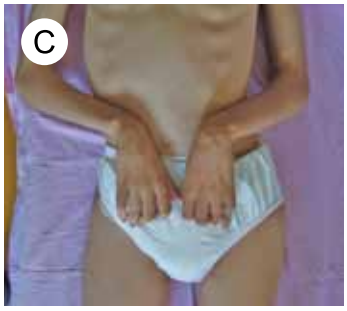
Sensory deficits, spasticity, loss of selective motor control, movement disorders such as chorea, dystonia and rigidity and muscle weakness are the reasons for upper extremity dysfunction in total body involved children. Visual and cognitive disability increase the problem. The child does not use the upper extremities and in time, develops contractures and deformities.

Many times there is no need for intervention beyond simple stretching and positioning. Functional splints may be useful. The shoulder internal rotation-adduction contracture does not interfere with function. The elbow flexion-pronation contracture creates problems when using forearm crutches [C]. Consider lengthening the spastic muscles and releasing the anterior capsule in a contracture of 100° and above to improve hygiene. Treat severe flexion contractures in the hand impairing hygiene and cosmesis with arthrodesis only after growth has stopped.

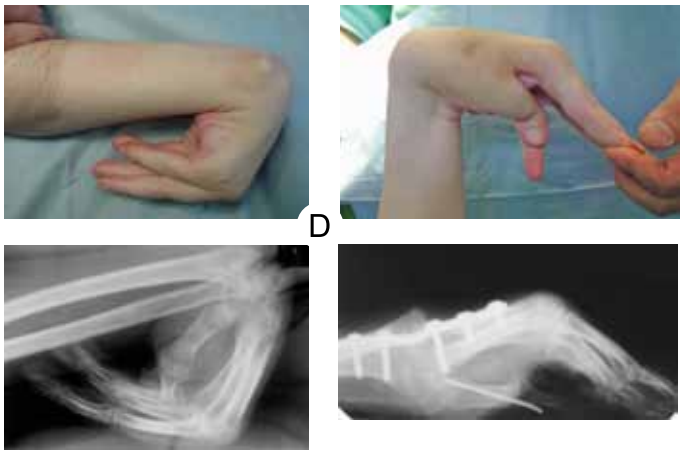
Knee flexion deformity prevents therapeutic ambulation.



Improper positioning results in equinovarus deformity.



Elbow flexion-pronation contracture and wrist flexion in a quadriplegic child impairs the ability to use the upper extremities for transfers.



Patient with a severe wrist flexion contracture was treated effectively with arthrodesis. Photos courtesy of G. Koloyan

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A

Movement problems in dyskinesia	
Athetosis	Involuntary, slow writhing movements of the hands feet face or tongue
Chorea	Multiple rapid jerky movements usually of the hands and feet.
Dystonia	Muscle tone is greatly increased. There are slow torsional contractions which increase with attempts at voluntary movement and result in abnormal posturing. Dystonia is localized more to the trunk and proximal extremities.

B

Classification	
Choreo-athetoid	*Dystonic*
Hyperkinetic	Rigid
Purposeless involuntary movements	Co-contraction of agonist & antagonists

Severe dystonia interfering with sitting and positioning may respond to medical treatment only.

Involuntary contraction of hand muscles prevents effective use of the extremity.

Dyskinesia

Athetosis, dystonia and chorea are the main movement disorders seen in dyskinetic children [A]. These children are initially hypotonic. As they get older, muscle tone begins to fluctuate. Involuntary movements occur when the child tries to move. Sometimes there is also movement at rest. When the child is totally relaxed in the supine position or asleep, there is full range of motion and decreased muscle tone. When the child wakes up or is excited, he becomes rigid. Lack of coordination is even more prominent during strenuous activities. The dyskinetic child spends excessive energy because of continuous uncontrolled movements. Abnormal contractions of many muscles occurring with the slightest voluntary motion, increase the energy demand considerably.

Classification

Dyskinetic patients are subdivided into two groups [B]. The first and most common group are hyperkinetic or choreo-athetoid children. They show purposeless, often massive involuntary movements. The initiation of a movement of one extremity leads to movement of other muscle groups. Rapid, random and jerky movements are called chorea and slow writhing movements are called athetosis. They increase when the child is excited or frightened.

The second group are dystonic children [C]. They manifest abnormal shifts of general muscle tone induced by movement. When the child tries to move, there is a co-contraction of agonist and antagonist muscles leading to an abnormal posture of one or more parts of the body. These abnormal and distorted postures occur in a stereotyped pattern. The trunk and neck are rigid. As in all types of dyskinetic CP, the contractions in the flexor and extensor muscles of the extremities increase with voluntary movement and disappear during sleep.

Dyskinesia may accompany spasticity in a group of total body involved children [D]. Athetosis is common in combination with spastic diplegia.

Associated features

Mental status is generally not impaired [E]. There is communication difficulty because of oromotor dysfunction and most of these children are unable to talk. Spasticity of oropharyngeal muscles impair feeding. Growth retardation and a decreased capacity to gain weight are characteristic.

Continuous repetitive muscle activity causes involuntary movements in all four extremities and the trunk in this 10 year old child. These movements increase when she wants to accomplish a certain task. Communication is difficult in spite of her normal mental function.

Musculoskeletal issues

The ambulation potential of dyskinetic children depends on the severity of involvement [A]. The majority of children with severe dystonia are unable to walk. Management is aimed at improving communication, independence in activities of daily living and wheelchair use. A percentage of children with athetosis can become ambulatory, however they have a clumsy and unstable gait. They lose their balance and fall easily when there is even the slightest disturbance from the world surrounding them. Contractures are almost never seen. Degenerative hip disease and acetabular dislocation are common complications during the adolescent growth spurt, particularly in children with athetoid cerebral palsy. Scoliosis is common. Complication rate of spine surgery is high. Cervical spine fusion is an option for treatment of advanced degenerative disease of the spine and C5-6 instability in the adult.

Treatment

Children with dyskinetic CP do not benefit from medical treatment, physiotherapy or orthopaedic surgery. Medical treatment options are many in dyskinesia, however their efficacy is questionable. The aim is to minimise muscle contractions and unwanted movements, to ease the burden of care, and to lessen the child's discomfort [B]. The use of intrathecal baclofen pumps are becoming increasingly popular in dystonic children.

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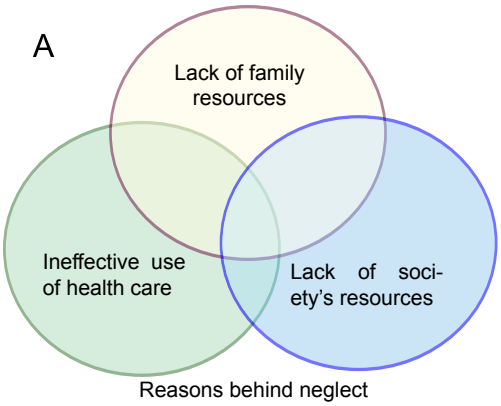
This child is able to walk with a walker but prefers crawling because dystonia disturbs her balance.

23 year old mixed CP with severe dyskinesia and spasticity. His face, neck trunk and extremity muscles are all affected. He cannot sit in the wheelchair because of forceful muscle contractions. Intrathecal baclofen pump implantation may be a possible treatment alternative, however keeping the catheter in place would be a major challenge in such a severe case.

A



B	General consequences of neglect
	Secondary mental deprivation
	Failure to thrive
	Social isolation
	Loss of mobility and ambulation
	Increased burden on the family
	Early mortality

C

Neglected 21 year old woman before and after treatment: Prior to surgery, she had severe knee flexion and ankle plantar flexion contractures, she had poor sitting balance. Simple hamstring muscle and Achilles tendon lengthenings were sufficient to improve sitting balance and enable therapeutic ambulation in solid plastic KAFOs.

The Neglected Child

Some children with CP cannot receive proper medical care because of reasons related to the family, the society and to the health care system they live in [A]. Lack of proper care by the family is one of the important reasons of neglect. The parents lack financial resources or are psychologically unable to provide adequate care for their disabled children. Families who are initially hopeful try to keep up with the demands of caring for a disabled child. They are frustrated or disappointed if the their child does not achieve what they expect. Eventually they stop providing even the basic treatments such as home exercises because they think that their efforts are futile. Some families are ashamed of having a disabled child for cultural reasons.

In certain parts of the world the society is not well prepared or does not have the resources to accept and live with the disabled. The community is not organized to continue the care of the child with CP at school or at home. Opportunities for special education, recreation, vocational training and sheltered work are extremely limited. The child who cannot use a wheelchair outside the house because of environmental barriers remains confined to the house and loses skills. The adolescent or young adult with CP who cannot find a job has no reason to leave the house so he loses his ambulatory skills.

The needs for disabled people remain unmet worldwide, and particularly in low/middle income countries where health service infrastructure, education and public awareness are lacking. Resources for health care and medical education are limited in many countries around the world. These limited resources are often not used effectively because of a lack of information. The information on CP that is available is often incorrect, out-dated and sometimes even promotes harmful treatments. Physicians and other health care providers lack up-to-date education in the treatment of CP.

No matter what the reasons behind the neglect are, neglected children are unable to reach their full potential and become a burden for their caregivers in the long run. The child with diplegic CP is hurt most by neglect because he has a great potential that is wasted [B].

Physicians treating CP patients meet such patients from time to time when the families decide to provide medical care for their children at some point in their lives or when charity organizations decide to finance treatment efforts. Most neglected children need orthopaedic surgery for better function [C]. The decision to perform surgery is risky because these children have been neglected for a long time and prognosis may be poorer than expected. The child and the family may not comply with the necessary prolonged and intensive postoperative rehabilitation. The child's medical and psychosocial status may not allow major interventions. There are basic clues to making decisions about treatment of the neglected child that spring mainly from experience. A 'family-centered' approach in delivering health services is likely to improve the wellbeing of children and their parents, and parental wellbeing is believed to be associated with improved outcomes for the child. The physician alone has to cope with this burden and assume especially the roles of the therapist, psychologist and social worker in countries where multi-interdisciplinary care is lacking.

Consequences of neglect are different for diplegic and total body involved children [A on next page].

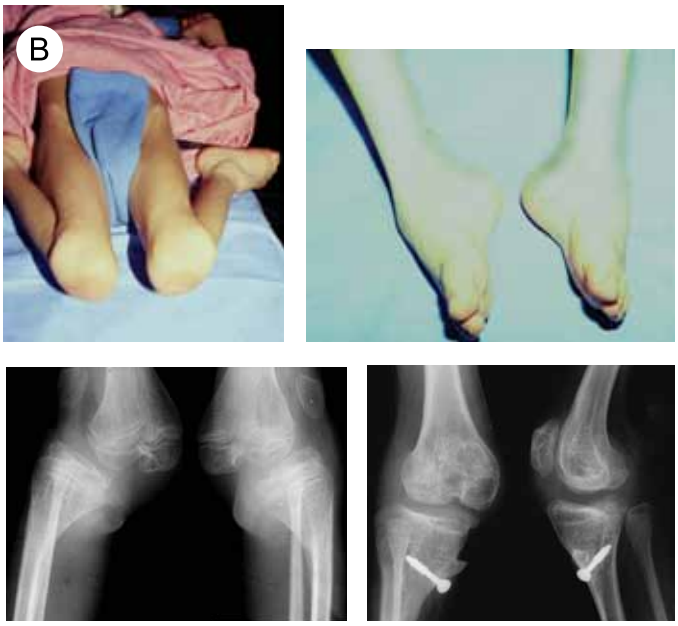
The total body involved child

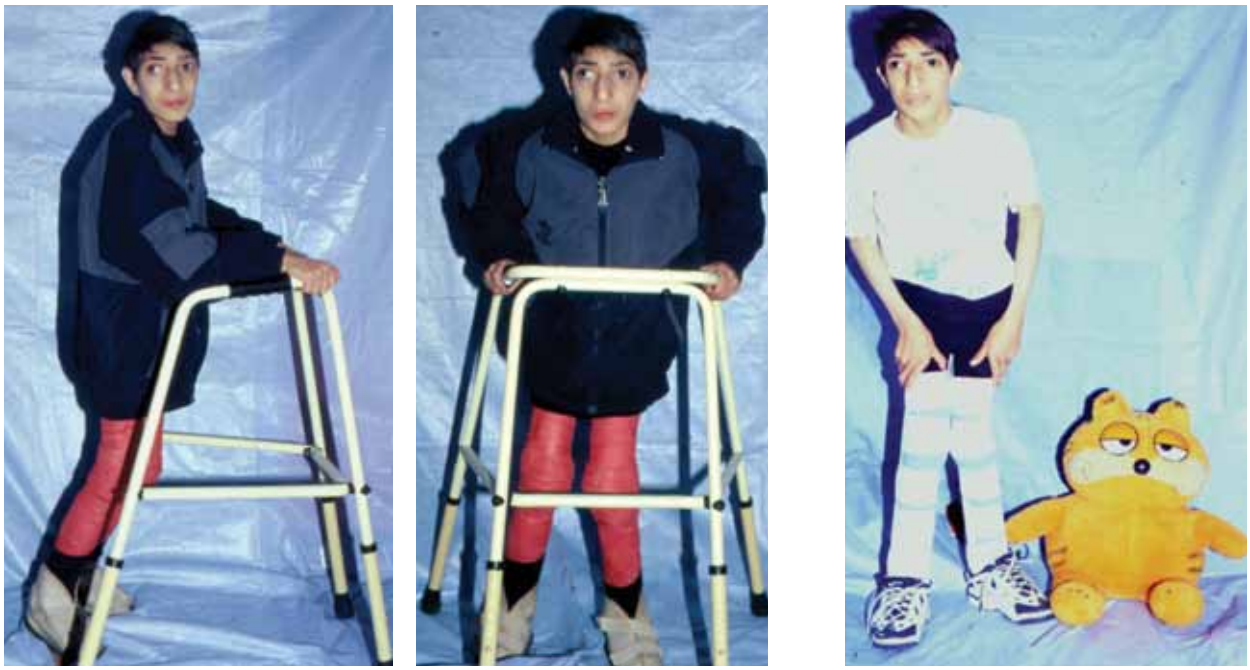
The main problems of the neglected total body involved adolescents and adults are spinal deformity and painful hips interfering with sitting as well as knee and ankle flexion contractures which prevent transfers. The patients also have severe hand flexion deformities. Growth disturbance, frequent infections and poor nutritional status almost always accompany the movement problem. Spasticity and dyskinesia are another major concern. Define the expectations clearly and get the parents' consent before advancing with treatment procedures.

Spine surgery is a difficult operation that places a great burden on the family and the child. Morbidity and mortality risks are high because of the poor general medical condition. Consider spine surgery only if there is a strong family support even if the patient's medical condition permits. Proper preoperative care does not decrease the risk of complications after spine surgery.

Operations for the painful hip are relatively easy, but families prefer nonsurgical intervention most of the time. Advise analgesic medications and proper positioning. Perform hamstring and Achilles tendon lengthenings if there is a potential for standing and therapeutic ambulation. Do not attempt temporary measures such as phenol or botulinum

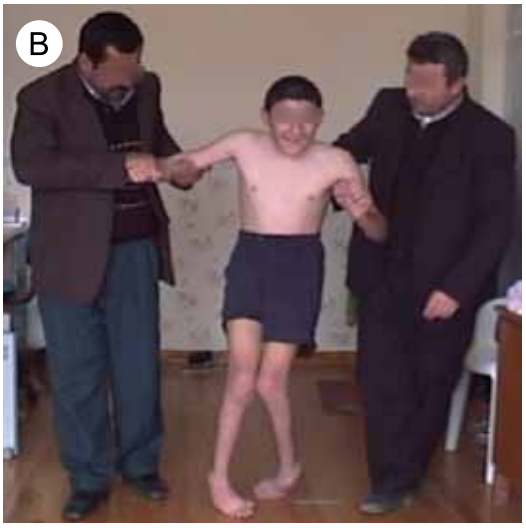
Consequences of neglect	
Total body involved	*Diplegic and hemiplegic*
Failure to grow	Loss of motivation to move
Poor nutrition	Fear of falling
Frequent infections	Fragile bones
Painful hips	Painful knees
Severe spinal deformity	Severe knee flexion
Knee and ankle contracture	Ankle plantar flexion





Severe patella alta, knee flexion and equinus contractures in a neglected adolescent were treated with osteotomies and advancement of tuberositas tibia. The child was able to use a walker and stand in plastic KAFOs after surgery. Courtesy of G. Koloyan

Neglected 11 year old diplegic. He can crawl around the house but the equinus deformity is a major problem when he tries to stand.



Neglected 16 years old boy with spastic diplegia. He has severe flexion contractures of both lower extremities, femoral anteversion and pes equinovarus. He can walk a couple of steps with the assistance of two people. He has never used a walker and never received treatment of any kind.

toxin injections in this group of patients who seek more radical solutions to their problems [B].

The diplegic child

The neglected diplegic child is probably the saddest situation that physicians treating children with CP will encounter. Most of these children have the potential to walk, but have been confined to immobility because of neglect [A]. Common problems include multiple severe deformities of the lower extremities. Hip problems are uncommon, instead, knee pain is present because of degenerative changes and overuse because of crawling on the knees. Children learn to walk between the ages of 4 to 7. It becomes difficult to teach them once they have missed that period in their lives. As the child grows older he loses the motivation to move, starts to feel afraid of falling and hurting himself. Bones are fragile and not used to carrying the body weight. The elderly immobile child has learnt to receive what he wants to have without spending any effort to move.

The neglected diplegic needs bone surgery as well as muscle tendon lengthenings to correct his deformities and to enable him to stand in an erect posture. Muscle weakness, bone pain and loss of selective motor control are much more pronounced compared to the young child who received adequate therapy. All deformities can be corrected, but correction of deformity does not always result in functional gain [B].

The postoperative rehabilitation period is tiring both for the child and the treatment team. Pain is an important obstacle to mobilisation. There is need for aggressive analgesic treatment. Fractures may be seen with intensive exercises. Intravenous bisphosphonate may prevent fractures. Bracing is difficult because of increased spasms and also decreased skin tolerance. It is difficult to gain ambulation in a child who has been in a wheelchair for a couple of years especially if that child does not have a social life outside the house that gives him/her incentive to walk. In spite of all, children who have good intelligence and strong motivation should be given the chance of ambulation through orthopaedic surgery and aggressive rehabilitation.

The hemiplegic child

The problems of the hemiplegic child are rather mild compared to total body involved or diplegic children. They become functional adults even if they do not receive physiotherapy, bracing or spasticity treatment in early childhood. The problems they will encounter are flexion contractures of the hand and equinus contracture of the foot. Hand surgery generally does not result in functional gains because of poor sensation and neglect. Equinus contracture will respond to Achilles tendon lengthenings. The patients do not like to use AFOs after surgery especially if they have been used to walking tiptoe for a long time.

The Adult

Thanks to increased awareness of the community integration of disabled people, more children with CP [A] are becoming adult members of the society. Despite the fact that adult CP patients [B] continue to have similar problems they had as children, they often do not receive adequate medical care and physiotherapy. Diplegic and hemiplegic adults have normal longevity. Both hearing and vision become worse with age. Total body involved adults continue to have the problems they had as children, namely; seizures, drooling, feeding and dental issues. 9% to 10.5% of adult patients with cerebral palsy have cardiovascular problems, including arterial hypertension and coronary artery disease. The goals of management and the modalities [C,D] remain the same though aging affects the outcome of treatment [E].

There are certain aspects where the adult CP patient is different from the child. Some of the special problems of the adult are pain, increased rate of fractures, scoliosis and dietary issues.

The halo effect: As the baby with CP grows and becomes an adult he loses all the sweetness and cuteness of infancy and childhood. He gradually turns into a disabled adult and the people around him stop treating him with the affection and sympathy they had when he was a cute little child. This change in attitude is difficult to handle and the adult with CP is pushed towards social isolation.

Goals of management
Maintain function
Maintain walking
Treat pain

C

Management modalities
Physiotherapy
Analgesic medication
Antispastic medication
Orthopaedic surgery

D

E	Effects of aging on outcome of therapy
	Prominent muscle weakness
	More time and effort for strengthening
	Less cardiovascular capacity
	Slower recovery

General problems of the adult	F
Musculoskeletal pain	
Neck 50% in spastic, 75% in dyskinetic CP	
Back	
Hip	
Knee	
Foot	
Contractures	
Overuse syndromes (in wheelchair or assistive device users)	
Fractures (more common in ambulators)	
Scoliosis (more common in nonambulatory patients)	
Gastrointestinal problems	
Constipation	
Reflux	
Dental problems	
Drooling	

Special problems of the adult patient

Pain

Pain in the nonverbal patient is difficult to understand and evaluate. The patient is agitated, restless, does not eat or sleep well. Perform an extensive work-up to determine the cause of pain. Differential diagnosis includes musculoskeletal problems, gastroesophageal reflux leading to ulcers, urinary or gynaecological problems and menstruation. Common musculoskeletal system problems causing pain in the nonambulatory adult are cervical spine degeneration, scoliosis and hip pathology. Common musculoskeletal system problems causing pain in the ambulatory adult are hip, knee and foot deformities. Physiotherapy and simple analgesics may help. Consider surgery in severe cases.

Fractures

Adult quadriplegic CP patients have osteopenia. They have a lower dietary intake of calcium. Decreased exposure to sunlight, immobility, spasticity, and the metabolic conversion of the precursors of vitamin D to inactive metabolites by anticonvulsant medications predispose the patients to fractures. Osteoporosis becomes worse as the patient ages.

Scoliosis

Scoliosis occurs in 25% to 64% of institutionalized adults. Uncorrected scoliosis may result in decreased ambulation and decubiti.

Sexuality issues

Adolescents with cerebral palsy have delayed and prolonged puberty. The reason is poor nutritional state. They may develop precocious puberty as well. Try and recognize the timing of sexual maturation and provide age-appropriate sexual education. Also try and determine if the patient is sexually active. Pose questions regarding sexuality privately, using normalizing statements and open-ended questions.

Feeding and nutrition

Feeding problems in adolescents with low caloric intake may result in poor growth and decreased muscle mass at maturity. They result in an adult with low fat-free mass. Athetoid patients have higher caloric requirements. Reductions in appetite and weight are harmful to the adult who already has a low fat-free mass and resultant malnutrition. A diet with sufficient iron (particularly in female patients) is important, because iron deficiency anemia is common in women with cerebral palsy.

Orthopaedic surgery in adult CP
1. Muscle releases lengthenings and transfers for contractures
Hip flexor /adductor
Hamstring
Gastrocnemius-soleus
Rectus femoris
2. Spine surgery for
Scoliosis
Back pain
Neck pain
3. Hip surgery
Total hip replacement
Valgus osteotomy
Resection arthroplasty
5. Bone surgery
Femoral or tibial derotation osteotomy
Triple arthrodesis
Hallux valgus surgery

A

B

Problems of the ambulatory adult
Deterioration of walking Greater energy expenditure when walking Less exercise No physiotherapy
Psychosocial problems Depression Social isolation
Musculoskeletal problems Subluxated painful hips Malalignment syndrome Patella alta and knee pain Pes valgus-hallux valgus

C

This adult patient has multiple lower extremity deformities but there was no need to intervene because he has an efficient gait and functions well in the society.

General goals of management

The goals of management in the adult with CP are to maintain function, maintain walking and to prevent or treat pain. Physiotherapy, analgesic and antispastic medication and orthopaedic surgery [A] have definite roles in this patient group. Oral tizanidin, diazepam or baclofen are options for spasticity treatment. The intrathecal use of baclofen is another alternative. Aging affects the outcome of all therapy procedures. Muscle weakness is more prominent in the adult compared to young children. Strengthening takes almost twice as much effort and energy. Cardiovascular capacity of disabled adults is markedly less than able bodied individuals. Recovery process after surgery is much slower.

The ambulatory patient

Deterioration of walking is the most important issue in ambulatory diplegics [B]. Adult diplegics have a greater energy expenditure when walking, because of their bigger and heavier bodies. They exercise less, and receive almost no physiotherapy [C]. Depression is a problem in the adult patient. They lose the family support they had as a child and become socially isolated. Social isolation and depression contribute to the deterioration in walking ability.

Because of a lack of exercise rate of contractures may increase. Release and lengthen the involved muscles to treat flexion and/or adduction contracture of the hip. Provide intensive post-operative rehabilitation. Hamstring tightness causes crouched gait, short stride length and kyphosis when sitting. Lengthen the muscles to relieve this problem. Heel cord tightness and valgus/varus deformities of the feet respond to lengthening, muscle releases and split transfers.

Special problems encountered in the ambulatory adult CP patients are subluxated hips causing pain, and malalignment syndrome causing painful knees and foot deformities. Hip subluxation is rare in the ambulatory CP child, but hip pain because of subluxated or dislocated hips may be seen in the adult. Treatment of choice is total hip arthroplasty. Apply hip spica casts for three weeks after total hip replacements to prevent early dislocations and relieve pain. Encourage the patients to stand fully weight bearing in the cast.

Spastic rectus femoris working against tight hamstrings causes patella alta and leads to knee pain. Consider distal rectus femoris and intermedius tenotomy combined with distal hamstring lengthening. Osteoarthritis of the knee is rare.

Another important problem of gait in the ambulatory adult is the malalignment syndrome presenting as a combination of femoral anteversion and external tibial torsion [A on next page]. Malalignment syndrome results in patellofemoral osteoarthritis and painful knees. Treat with proximal femoral derotation and supramalleolar rotation osteotomy.

Common foot deformities are bunions (hallux valgus), claw toes and severe pes valgus. The standard procedure of metatarsophalangeal fusion is performed for hallux valgus. Consider resection arthroplasty, proximal interphalangeal fusion or the Ruiz procedure for claw toes. Severe pes valgus is usually associated with external tibial torsion. A treatment option is supramalleolar rotation osteotomy with triple arthrodesis [B on next page].

The nonambulatory patient

Adults are physically bigger, therefore the care and transfer of the adult total body involved patient becomes a burden for the caregiver. Non-ambulatory adults often have severe osteoporosis with an increased rate of fractures. Wheelchair accommodations are sufficient for contractures that do not interfere with sitting or standing transfers in nonambulatory adults. Special problems of the nonambulatory adult are scoliosis, lack of hip abduction and knee pain [C].

Scoliosis can be progressive even in adults. Consider extensive spinal fusion if contoured wheelchairs or TLSO braces are not sufficient to provide adequate sitting balance.

Lack of hip abduction causes difficulty with hygiene and sitting. The cause of hip pain in the adult is hip subluxation and dislocation [D]. Simple analgesics and physiotherapy may be helpful. Total hip replacement is becoming increasingly popular because it offers the advantages of stability and standing for transfers [E]. Resection arthroplasty [F], arthrodesis [G] or valgus osteotomy [H] are other options.

CP is not just a pediatric problem. Exercise, stretching and other management modalities are lifelong commitments. Physicians and therapists alike need to be well prepared to deal with the problems of the adults with CP.

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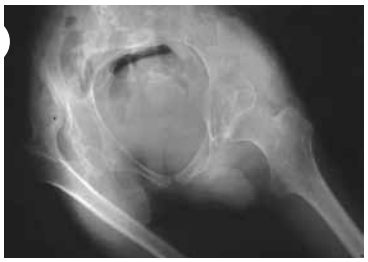
Miserable malalignment characterized by femoral anteversion, tibial external rotation and planovalgus.

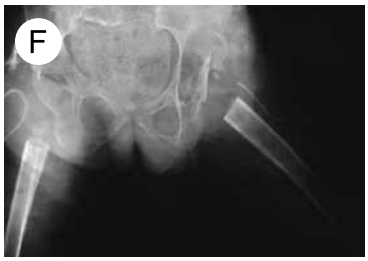
This young man is a severely involved mixed diplegic. Dystonia and ataxia limit his walking capacity. He has pes valgus and spontaneous extension in both great toes.

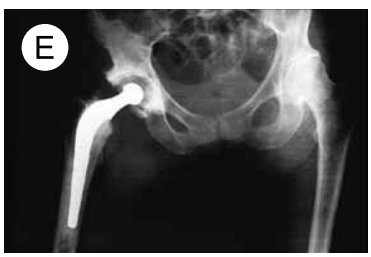
Problems of the nonambulatory adult

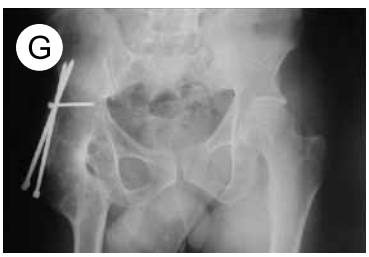
- Problems with care and transfer
  - heavier
  - severe contractures
  - lack of hip abduction
- Fractures
  - Osteoporosis
- Hip pain
  - Subluxation
  - Dislocation
- Scoliosis

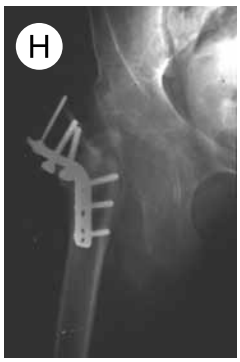
C











Treatment options for the painful hip of ambulatory adults are total hip prosthesis, resection arthroplasty, arthrodesis and valgus osteotomy.

Management With Limited Resources

CP is a worldwide problem. It spares no country or geographical location. The incidence of babies born with CP is the same around the world, however the prevalences at the time of school entry are different. This may mean that some children are lost by the time they approach school age or that some never have the chance to enter any sort of education.

There are many options for managing the child with CP to make him part of the society, to improve his quality of life as well as help his family. Even in well developed countries resources are rich but not infinite. In most other parts of the world disabled children are not lucky enough to benefit from most advanced technological improvements such as powered wheelchairs or newly developed drugs such as botulinum toxin. There is a limitation of specialized medical staff, equipment and finance. It remains to the physician to use his skill to help these children. The success of treatment depends upon an effective use of resources of the family, society and the health care system.

The principles of management with limited resources are to use the least expensive, time consuming and relatively more effective methods to deal with the problems of these children and to enable them to use the existing educational and vocational resources of the community they live in. In this context, the question of what is necessary and what is a luxury becomes a major concern.

What happens when resources are limited?

Hemiplegia

Almost all children who have hemiplegic CP can become independent adults. They may have contractures and deformities but function efficiently despite these. Some with seizures, learning disabilities and behavioural problems experience difficulty attending school.

Diplegia

Most diplegic children have the potential to walk. They benefit a lot from all treatments to decrease spasticity and to improve walking capacity. When resources are limited they cannot fulfil their potential and remain nonambulatory or floorbound for mobility. Mobility is directly related to integration into the society and independent living in most parts of the world. In countries where health care resources are limited education opportunities are also limited and children with impaired mobility have a greatly decreased chance of getting a proper education.

Quadriplegia

Quadriplegic children cannot be independent and need continuous care. They need proper health care and adequate nutrition to survive beyond adolescence. They also benefit from treatments to decrease spasticity and from equipment for sitting, mobility, communication and education. When resources are limited, a higher percentage of children die early. The survivors and their families have poor life quality.

What to do when resources are limited?

In most countries the medical treatment of children with CP is the responsibility of the family rather than the state. Therefore it becomes very important that each penny spent for treatment gets good return. Keep this in mind and select the treatment that is scientifically proven to be valuable.

Try to make the child as independent as possible for a better future. Special education can be very important in this regards. Tell the parents that physiotherapy improves only motor component of the child. Have them spend time for communication, cognition, self help and social development.

Provide a home bound program for children coming from far away places. Address the basic needs of the child and the family. Provide the opportunities for the child to get an education. Teach the family basic exercises to prevent contractures and deformities. Try and increase the level of communication. Find a way to establish a useful purpose for the child in the society so that he will be integrated. Aim to involve all the family members into caring for the child. Get support from the brothers and sisters of the disabled child.

The necessities

For all children the basic treatment should include positioning, stretching and strengthening exercises.

Children with walking potential

Simple solid AFOs are necessary to improve walking in the ambulatory children and to prevent contracture in the child who sits in the wheelchair.

Children without walking potential

Severely involved children need abductor pillows to prevent hip instability. They may need KAFOs for therapeutic ambulation. KAFOs at rest and at night may help prevent hamstring contractures. Severely involved total body involved children need proper seating arrangements in a wheelchair. A TLSO strapped to the wheelchair will provide the necessary trunk support. Oral antispastic agents such as baclofen and diazepam are readily available in many countries around the world, they are cheap and relatively safe. Gastrocnemius, hamstring and adductor lengthening surgery are safe, easy and reliable procedures to relieve spasticity and improve walking in ambulatory children. Progressive hip instability is a major problem which impairs the life quality of the child, decreases survival and increases caregiver burden. Early adductor and psoas tendon releases may help prevent hip subluxation.in the presence of hip flexion and adduction contractures. If subluxation exists however, soft tissue releases alone will not be helpful.

The child who cannot communicate but has normal mental function can easily use a communication board which contains a set of pictures or symbols. Simple methods to provide basic educational needs exist and can be taught to mothers. Feeding and constipation problems may be solved using a daily routine and feeding the child at regular short intervals with food in liquid form.

Improving mobility is the most important issue worldwide. For the total body involved child, a manual wheelchair driven by caregivers may be the basic option. Powered children's wheelchairs may be unavailable or too expensive for certain parts of the world. Unfortunately in many regions, environmental barriers limit the use of powered wheelchairs.

Advice For Families *

What is cerebral palsy (CP)?

Cerebral palsy is a medical condition that affects control of the muscles [A]. It appears in the first few years of life. Cerebral means the brain's two halves, and palsy means inability to move properly. Cerebral palsy is not a problem of the muscles or nerves. Instead, it is a problem with the brain's ability to adequately control movement and posture.

What happens if someone has cerebral palsy?

If someone has cerebral palsy they are not able to use some of the muscles in their body in the normal way because of an injury to their brain. Children who have cerebral palsy may not be able to walk, talk, eat or play in the same ways as most other kids. They have difficulty with fine motor tasks, such as writing or cutting with scissors; experience trouble with maintaining balance and walking; or be affected by involuntary movements, such as uncontrollable writhing motion of the hands or drooling.

The severity of the problem is different in every child. Some have very mild problems whereas others have very severe involvement and other medical disorders, including seizures or mental impairment [B].

Why does it occur?

CP is caused by an injury to the brain before, during, or shortly after birth. In many cases, no one knows for sure what caused the brain injury or what may have been done to prevent the injury.Brain damage in the first few months or years of life can follow head injury or brain infections, such as bacterial meningitis or viral encephalitis.

The cause of brain damage before or during birth is unknown. Risk factors that increase the likelihood of brain damage are prematurity, low birth weight and difficulties during pregnancy. Doctors should keep an eye on children who have these risk factors [C].

In the past, doctors thought that cerebral palsy occurred because of asphyxia or hypoxia during birth. However, research has shown that very few babies who have birth asphyxia develop CP. Birth complications are now estimated to account for about 6 percent of cases.

CP is not contagious and it is not inherited from one generation to the next.

How common is CP?

CP occurs in 2 of every 1000 babies worldwide. The United Cerebral Palsy Associations estimate that more than 500,000 Americans have cerebral palsy. Despite the many technological advances in medicine, the number of children with CP remained the same over the past 30 years. This is partly because more critically premature infants are surviving through improved intensive care. Unfortunately, many of these infants have nervous system damage.

What are the early signs?

Signs of CP appear before 3 years of age [D]. Parents first suspect that their infant is not developing normally. Infants with cerebral palsy are slow to learn to roll over, sit, crawl, smile, or walk. This is called developmental delay.

Some children have hypotonia when the baby may seem flaccid and relaxed, even floppy. Some have hypertonia, and the baby seems stiff or rigid. In some cases, the baby has an early period of hypotonia that progresses to hypertonia after the first 2 to 3 months of life. Affected children may also have unusual posture or favour one side of their body.

A

A damage to the centers that control movement in the developing brain results in cerebral palsy. The child has difficulty moving and maintaining his balance.

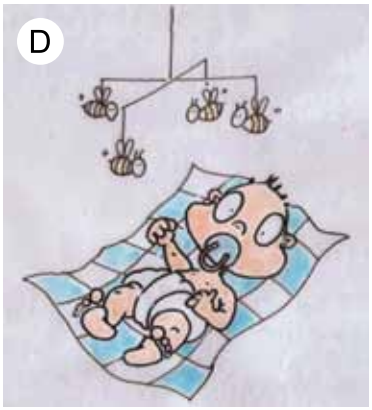
B

Children with CP may have:
Tight and stiff muscles called spasticity
Muscle weakness
Balance problems
Involuntary movements
Mental problems
Attention and perception deficits
Seizures
Visual problems
Hearing problems
Communication problems
Poor nutrition and failure to gain weight and grow
Drooling
Dental caries

C

Risk factors
Breech presentation
Complicated labor and delivery
Low Apgar score
Low birth weight and premature birth
Multiple births
Nervous system malformations
Maternal bleeding or severe proteinuria late in pregnancy
Maternal hyperthyroidism, mental retardation, or seizures
Seizures in the newborn

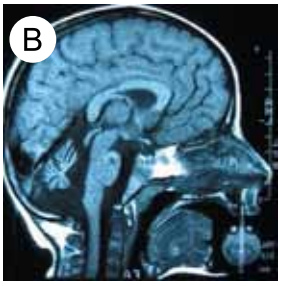
D



The early signs of CP are apparent from 2 months of age. The baby is unable to hold the head erect, he lies in an asymmetric posture and favours one hand. He is irritable or extremely quiet and does not suck well.

** Illustrations by Idil Çilingiroğlu*

The diagnosis is made by clinical examination and accurate history.



Cranial MRIs can reveal the brain abnormality from 2 months onwards in many cases. The baby has to hold still for at least 20 minutes so there is a need for anesthesia. Repeat MRIs are generally not necessary.

Quadriplegic children may also have dystonia in his hands.

Ataxic children can walk if they can hold onto their caregivers or a walker.

A physician should be consulted when there is concern about a baby's development [A]. The physician can distinguish normal variation from a developmental disorder.

How is it diagnosed?

CP is diagnosed by history and clinical examination. Slow development, abnormal muscle tone, and unusual posture indicate CP. The physician must determine that the child's condition is not getting worse. CP is not progressive. If a child is continuously getting worse, he probably does not have CP.

There are specialized tests to learn more about the possible cause of cerebral palsy. Computed tomography (CT), uses X rays and a computer to visualize the brain tissue. A CT scan shows abnormal brain areas. Magnetic resonance imaging (MRI) [B] uses a magnetic field and radio waves. It shows the brain lesion more clearly in certain cases. These scans do not prove whether a child has a cerebral palsy, and they do not predict how a specific child will function as she grows.

Ultrasonography bounces sound waves off the brain and uses these to view the brain structures. It can be used in infants before the bones of the skull close. It shows the abnormality in the brain, is less expensive, and does not require long periods of immobility.

An electroencephalogram, or EEG will show the natural electrical currents inside the brain and will reveal a seizure disorder if present.

Intelligence tests are used to determine if a child with CP is mentally impaired. An ophtalmologist and an otologist are necessary for vision and hearing problems.

Once the child is diagnosed with CP there is no more need for repeated MRI scans.

Are there different types of CP?

Children with CP have damage to the area of their brain that controls movements. Their problems are: muscle weakness, tight and stiff muscles, balance problems and coordination difficulty. The muscles are either too tight, too loose, or a combination of tight and loose.

Spastic CP

Spastic means that the muscles are too tight. Children with spastic CP have stiff and jerky movements because their muscles are too tight. They often have a hard time moving from one position to another or letting go of something in their hand [C]. This is the most common type of CP.

Dyskinetic (athetoid or dystonic) CP

Children with dyskinetic CP have trouble holding themselves in an upright, steady position for sitting or walking and often show lots of movements of their face, arms and upper body that they don't mean to make (random, involuntary movements). These movements are usually big and increase when children are excited or frightened. During sleep they go away.

Children may not be able to hold onto things (like a toothbrush or fork or pencil) because of their mixed tone and trouble keeping a position.

Ataxic CP

The muscles are loose and the child has difficulty maintaining balance and coordinating his movements. Kids with ataxic CP look very unsteady and shaky. They shake a lot especially when they are trying to do something. They have poor balance and walk unsteadily [D].

Mixed CP

When spasticity, ataxia and dyskinesia occur together in the same child, we call it a mixed type CP.

Besides different kinds of muscle tone, we must also define which parts of the child's body is affected. This depends on the extent of the brain injury.

Quadriplegia

When a child has a movement problem in all four of his limbs, it is called quadriplegia. These children have trouble moving all the parts of their bodies, their face and trunk [A]. They use a wheelchair to get around. They also have trouble talking and eating.

Hemiplegia

When a child has movement problem in one side of the body only, it is called hemiplegia. The other side of the child's body works fine. Many children with hemiplegia are able to walk and run.

Diplegia

When a child has a movement problem just in the legs or much more severe in the legs than in the arms, it is called diplegia. Children have difficulty walking and running. They can hold themselves upright and use their arms and hands.

What other problems may be seen in children with CP?

Children with CP may have some other problems that are caused by the same brain injury. These include:

Mental impairment

About one-third of children who have cerebral palsy have mild, one-third moderate or severe mental impairments. The remaining third are normal. About one-fourth to one-half of children with CP also have learning problems [B]. They may have trouble with one or two subjects in school but learn other things well. They learn at a slower rate. Children with mild mental retardation learn to read and write and do math. Many children need some special learning help in school.

If a child does not have the means to move around he will be unable to explore his surroundings and interact with his peers [C]. This will result in secondary mental and social deprivation.

Seizures or epilepsy

About 30% of all children wih CP have seizures. This means some abnormal activity in their brains that interrupts what they are doing. Seizures usually last a few seconds to a few minutes, and are not dangerous. Many children take special medicine to help prevent seizures.

Growth problems

Children with spastic quadriparesis lag behind in growth and development despite having enough food. In babies, this lag usually takes the form of too little weight gain; in young children, it can appear as abnormal shortness; in teenagers, it may appear as a combination of shortness and lack of sexual development. The causes are poor nutrition and damage to the brain centers controlling growth and development [D].

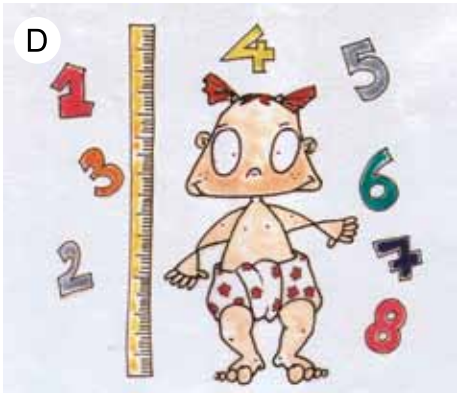
Feeding problems

CP can affect the way a child moves his mouth, face and head. The child may have difficulty biting, chewing and swallowing food, which can cause poor nutrition. Poor nutrition increases risk of infection and cause or aggravate the lag in growth and development.

The most important problem in CP is inability to move.

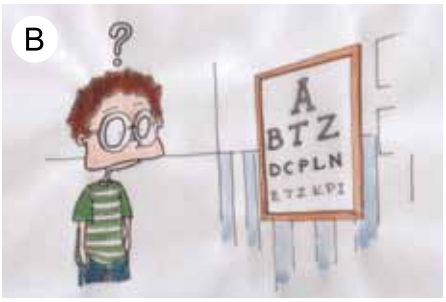
Children with CP may have learning difficulty.

The immobile child is isolated from his peers.



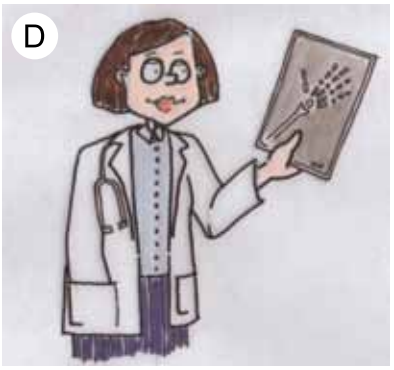
Growth is retarded because the child cannot eat properly. Adequate nutrition is essential for brain development in the first 3 years of life. Get proper evaluation if the child vomits all the time, cannot chew or swallow. A gastrostomy will help you feed your child.

Improved communication strengthens self-image. The child can have friends and socialize.



Poor eyesight increases the movement problem.

The diagnosis of CP can be made around the age of one by a pediatric neurologist.



A pediatric physiatrist designs a rehabilitation program for your child according to the child's needs and the treatment team's expectations

Prepare semisolid food, vegetables and fruits to make swallowing easier. Proper position and sitting up while eating or drinking is also helpful. In severe cases doctors recommend a gastrostomy in which a tube is placed directly into the stomach.

Communication

The child may not be able to control her lips, jaw and tongue much. He also has trouble controlling his breath flow to make his voice work. Therefore he has difficulty talking clearly and making himself understood [A].

Drooling

Children with poor control of mouth and pharynx muscles have drooling. This can cause severe skin irritation and also lead to further isolation of affected children from their peers. Drugs can reduce the flow of saliva but may cause mouth dryness and poor digestion.

Incontinence

This is caused by faulty control over the muscles that keep the bladder closed. The child may wet the bed, leak urine during physical activities or spontaneously.

Impaired vision or hearing

Strabismus occurs in a large number of children. This is a condition in which the eyes are not aligned because of differences in the left and right eye muscles. In children, the brain adapts to the condition by ignoring signals from one of the eyes. This leads to very poor vision in one eye. The child may be unable to judge distance. In some cases, surgery to correct strabismus may be necessary. Children with hemiparesis may have defective vision or blindness that impairs the normal field of vision of one eye [B]. Impaired hearing is also more frequent among those with cerebral palsy than in the general population.

Abnormal sensation and perception

Some children with cerebral palsy feel simple sensations like touch and pain less than normal. They may also have or difficulty identifying objects when their eyes are closed just by touching them.

Can it be cured?

The lesion in the brain cannot be cured, although the consequences can be minimized. Treatment can improve a child's capabilities. Many cerebral palsied people can live near normal lives. There is no standard therapy that works for all patients.

Who treats CP?

A team of health care professionals identify a child's needs and then to create an individual treatment plan for him. The members of the treatment team are knowledgeable professionals with a wide range of specialities.

The team includes

A physician (a pediatrician, a pediatric neurologist, or a pediatric physiatrist) trained to help developmentally disabled children [C,D]. This physician works to build a comprehensive treatment plan, implements treatments, and follows the patient's progress over a number of years.

An orthopedist who specializes in treating the child musculoskeletal system. An orthopedist diagnoses and treats muscle and bone problems associated with CP.

A physical therapist, who designs and implements special exercise programs to improve movement and strength.

An occupational therapist who helps patients learn daily living skills, also skills at school and work.

A speech and language pathologist who treats communication problems.

A psychologist who helps patients and their families cope with the special stresses of CP.

An educator who teaches children with mental impairment or learning disabilities.

The child with CP and his family are also members of the treatment team. As a family, be involved in all steps of planning, making decisions, and applying treatments.

Treatment plan includes drugs to control seizures and muscle spasms, special braces to make walking easier, orthopaedic surgery [A], mechanical aids to assist in daily life, counselling for emotional and psychological needs, and physical, occupational, speech, and behavioural therapy. Early treatment gives the child a better chance of learning to move.

Do not forget that the ultimate goal is to help the child have a happy life, healthy growth into adulthood and maximum independence in society [B].

Is there a drug to cure the problem?

There is no drug that can cure CP. Physicians usually prescribe drugs to stop seizures, to relax muscles and if necessary to stop drooling.

Diazepam acts as a general relaxant of the brain and body; baclofen blocks signals sent from the spinal cord to contract the muscles; and dantrolene interferes with the process of muscle contraction. They are effective for short periods and have side effects such as drowsiness. The long-term effects of these drugs on the developing nervous system are unknown. Patients with dyskinetic cerebral palsy may need drugs that reduce abnormal movements.

What is botulinum toxin? Is it useful in CP?

Botulinum toxin is a drug that decreases spasticity when injected into the muscle. It stops the signal for contraction from the nerve to the muscle. Children who walk on tiptoe or with bent knees benefit from botulinum toxin injections [C]. The physician decides whether botulinum toxin is suitable for your child and does the injections. The injection is not painful but sedation is necessary. General anesthesia is better if many muscles are to be injected. The effects starts at 3-10 days and continues for 3-6 months. Re-injections can be done if necessary. After the injection, the physician may decide to use a short or long leg cast. Physiotherapy is essential. Wearing braces is easier, the child walks better and has better balance. Relieving spasticity in this manner may preserve muscle length and minimize the need for orthopaedic surgery. There are almost no side-effects. A slight weakness may be observed. Botulinum toxin is not useful in generalized spasticity.

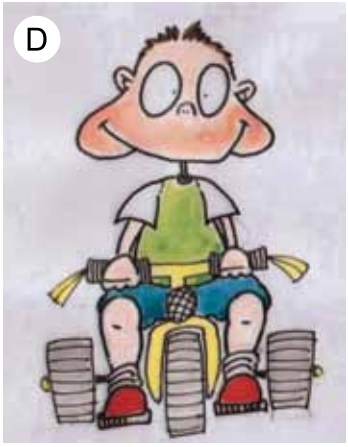
Are physical therapy and physiotherapy the same? How much does my child need?

Physical therapy and physiotherapy are different names for the same therapy method. Physiotherapy involves treatment of the musculoskeletal system with exercises to regain joint movement, muscle strength and mobility. Physiotherapists try to teach the child how to move better and how to maintain balance. At the same time, they also try to prevent the musculoskeletal system complications that occur because of muscle tightness and weakness. They teach children with CP to walk, use their wheelchair, stand by themselves, or go up and down stairs safely. Children must perform these exercises in fun activities like running, kicking and throwing a ball, or learning to ride a bike [D].

The pediatric orthopaedic surgeon treats the musculoskeletal problems that occur because of spasticity.

Education is the most important aspect of treatment. Every child with CP should get a proper education within the limits of his capacity. Even children who have mental retardation can learn.

Botulinum toxin injections relieve spasticity and do not cause harmful long lasting side effects.



The child must enjoy therapy. Exercises must be in the form of play activities. A tricycle is great for teaching reciprocal movement.

Strengthening is crucial for effective mobility. The child who has weak muscles cannot walk. Bicycles can be supplied with side wheels for balance.

Do not be afraid to use a wheelchair. It does not cause addiction. On the contrary, children who are mobilized at an early age with a wheelchair do not lose the motivation to move. Computer aided systems attached to the chair improve communication.

The basic brace in CP is the plastic ankle foot orthosis (AFO). Do not use KAFOs or metal braces.

Physical therapy begins in the first few years of life. The most common therapy technique is called the Bobath technique, named for a husband and wife team who pioneered this approach. Therapists try to provoke advanced control of movement by stimulating the child in this technique. There are also many other techniques used in various parts of the world like the Vojta therapy or the Rood method and others. All of these techniques have the same principles. One is not better than the other. It depends on the skill of the physiotherapist to use these various techniques in the best way to improve function. Do not worry about the therapy method your physiotherapist is using. Always concentrate on how functional and how happy your child is.

Strengthening exercises are necessary to prevent weakness and stretching exercises are essential to prevent contractures [A]. Normally, a growing child stretches his muscles and tendons during daily activities. As the child runs and plays muscles and bones grow together. Spasticity prevents this stretching in CP. As a result, muscles do not grow as fast as bones. Muscles get stiff and short, they prevent joint movement. This is called a contracture. Physical therapy works to prevent contractures by stretching spastic muscles. For example, if a child has spastic hamstrings (muscles in the thigh behind the knee), the therapist and parents should encourage the child to sit with the legs extended to stretch them.

Like all children, the child with CP needs to experience the world around him in order to learn. Stimulating the child with exercises and other therapy procedures can make this possible for the child who is physically unable to explore. Giving the child a wheeled mobility device is also very helpful.

As the child approaches school age, efforts to prepare the child for the classroom are necessary. Physical therapy can help the child prepare for the classroom by improving the ability to sit, move independently or in a wheelchair [B].

What braces can the child use?

It is important to mobilize the child. Use seating devices and head supports to enable better sitting. There are special devices called standers that help the child stand with support. These are necessary for more severely involved children. The most common braces are called AFOs [C]. The capitals stand for ankle foot orthoses. They are generally made of plastic from a plaster model of the child's foot. The physician will decide which braces are necessary and prescribe them for the orthotist to make. The KAFO (knee ankle foot orthoses) has been abandoned in CP as well as the metal uprights attached to orthopaedic shoes. Try and use the simplest and smallest brace so as not to interfere with walking. Do not use orthopaedic shoes, they are of no help.

Do children need speech therapy?

Speech therapy shows ways of communicating with the child. The child may learn to communicate through talking, using sign language, or using a communication aid. Children who are able to talk work with a speech therapist so that they will improve their speech for people to understand them better. They learn new words, to speak in sentences and improve their listening skills. Children who are not able to talk learn sign language or use a communication aid. A simple communication aid is a book or poster with pictures that show things the child might want, or an alphabet board that the he uses to spell out his message. There are also computers that actually talk for the child.

What is occupational therapy?

Occupational therapy is teaching the child activities of daily life. These include how to write, draw, cut with scissors, brush

teeth, dress [A] and feed or control the wheelchair. Occupational therapists help children find the correct equipment to make some jobs a little easier.

Is there time for play?

As a family, take time to have fun [B]. Do not let therapy take up all the child's time. The goal of therapy is to make the child have a normal childhood. By definition, this includes play activities. There are recreational therapists who work with children on sports skills or other leisure activities. Children may learn to dance, swim or ride a horse. They may also work on art or grow and take care of plants. Find out what your child is interested in and try to improve his capabilities [C].

Is surgery really necessary?

Orthopaedic surgery is necessary if there are contractures or if the hips are subluxating. The surgeon first determines the exact muscles that are spastic, because lengthening the wrong muscle could make the problem worse.

Lengthening a muscle makes it weaker. Children need intensive physiotherapy after surgery and recovery takes months. For this reason, doctors try to fix all of the affected muscles at once when it is possible.

No matter how well they are cared for, contractures and deformities occur eventually in all diplegic children. Therefore almost all diplegic children need orthopaedic surgery sometime in their lives.

A neurosurgical operation known as selective dorsal rhizotomy aims to reduce spasticity in the legs. Doctors try to locate and selectively cut overactivated nerves in the spine that control leg muscles. The results are not clear.

Are there any devices to make life easier?

Special machines and devices can help the child or adult with cerebral palsy overcome limitations. The computer makes the largest difference in the lives of those with cerebral palsy. A child who is unable to speak or write may be able to learn to control a computer using a special light pointer that attaches to a headband. Equipped with a computer and voice synthesizer, this child can communicate with others.

What is behavioural therapy?

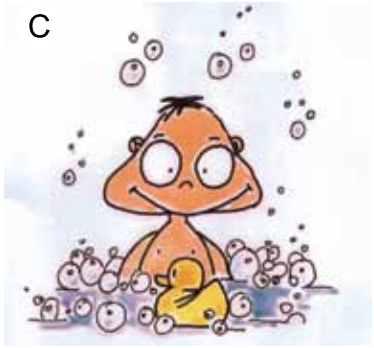
Behavioural therapy uses psychological theory and techniques. It might include hiding a toy inside a box to reward a child for learning to reach into the box with his weaker hand. Therapists may try to discourage unhelpful or destructive behaviors, such as hair-pulling or biting. As a child with cerebral palsy grows older, continuing physical therapy must be supplemented by vocational training, recreation and leisure programs, and special education when necessary. Counselling for emotional and psychological challenges may be needed at any age, but is often most critical during adolescence. Depending on their physical and intellectual abilities, adults may need attendant care, living accommodations, transportation, or employment opportunities.

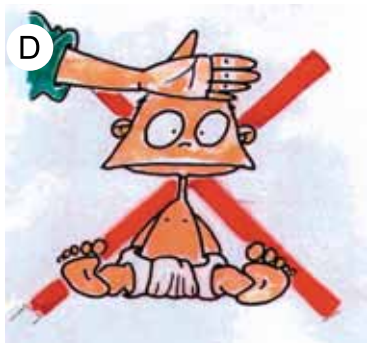
Can the child with CP walk? Will my child walk?

Predicting what a young child with cerebral palsy will be like or what he will or will not do is very difficult. It is only possible to a certain extent after the child is two years old. Children with cerebral palsy do not lose the skills they mastered. If a child loses a skill he previously could, look for a different cause of the child's problems [D].

Occupational therapy teaches the child daily living skills such as dressing and buttoning up.

A childhood full of fun and play is a happy childhood. It is possible even in CP.

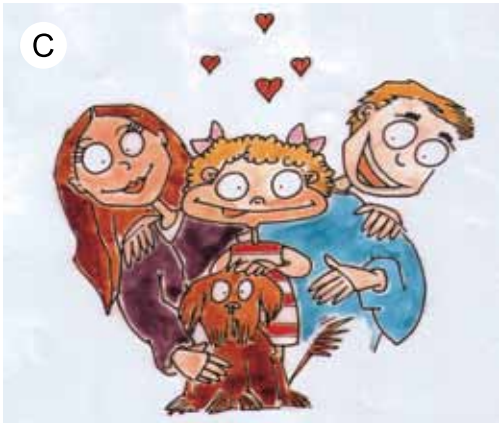




Walking is the most important priority of parents when the child is a baby, however, being able to walk is not important for happiness in an adolescent. Being loved, being able to give and receive, being productive makes a person happy. Teach your child to love and to be productive. Love your child so that he is happy.

Motivation to move and mental retardation are two most important requirements for ambulation and independent living.

All mentally capable children should attend normal schools regardless of the degree of their movement problem.



Maintain the integrity of your family. Spend time together, have fun together. Share good and bad experiences. Spare more time for each other than for therapy.

Parents should learn how to care for their baby at home. Holding the baby in certain positions during feeding, performing the exercises and playing with the baby stimulate the central nervous system.

In order for a child to be able to walk, he must be able to hold up his head and sit independently. A child who does not sit up by himself by age 4 will not walk independently.

Mental retardation impairs a child's ability to function. It is difficult to predict early on whether the child will be mentally retarded or whether he will be able to talk. Evaluating intellectual function is also hard because of the movement problem. Children who are mentally retarded have more difficulty walking. Children who do not have the motivation to move do not become independent ambulators despite adequate motor control [A].

CP does not always cause severe handicap. A quadriplegic child might be unable to walk and need extensive, lifelong care. A hemiplegic child is slightly awkward but requires no special assistance.

Can the child with CP go to school?

Yes, all hemiplegic children, mentally adequate diplegic and quadriplegic children must attend mainstream education. Prepare the teachers beforehand that the child with CP will need extra time in activities that require precise movements with the hands. A proper education is the only solution to the movement problem that limits the child's independence [B].

Can the child with CP live independently?

Independent living depends on many factors. The most important factor is mental retardation. The child must have an adequate intelligence to earn a living and support himself. Active use of the hand and functional mobility are also important factors to consider. Walking is important but not high priority. Most hemiplegic children but no quadriplegics can live independently.

Parents, keep in mind!

The joy of childhood

Your child is a child only once. Do not get carried away by treatments. Be proud of your child and let him discover the joy of childhood [C]. Have hope for, and become close to your child.

The joy of parenthood

Accept your child the way he is but try and get the best of treatment for him. Do not blame yourself, lose hope or give up. The cause of CP is unknown, it is not your fault, nor anybody else's. The diagnosis is difficult and there are no tests to prove whether a child has a cerebral palsy. No test can predict how a child will function as she grows.

You may feel tired, helpless and lonely. Sometimes you may blame yourself or think that you are neglecting your family and other children. These are normal feelings. Share them with loved ones and your physician. Do not ignore your own needs and your relations. You can help your child more if you are strong [D].

Forming alliances

Contact those families who have similar problems. Take part in associations and organizations. Be useful to others.

Improve your knowledge. Read all available resources and try to support the foundations for CP. Join the seminars, symposia and panels and follow up on new developments.

Parenting your child

Do not overprotect the child. Allow your child to explore his environment. Put his toys to a certain distance away from him and encourage him to get these himself. You can motivate him to move in this way.

Support your child to gain independent self care and daily life activities. He can wash his hands and face, put on and take off his clothes, eat his food or at least help you do these. Give him ample time.

Infancy When your child is just a baby, learn how to hold, feed and exercise him. Infancy is a time of uncertainties. Do not be discouraged and keep loving your baby.

Toddler When your child is a toddler support his body so that he can use his hands in active play. Encourage him to move by himself. Get him wheeled devices [A] so that he can move around the house and explore his surroundings. Never believe those who say the child should crawl before he can walk or that his spine will bend if you make him sit. See for yourself what your child is capable of doing and support him if he wants to do that.

Schooling Never neglect the child's education. Defend your child's right to get the best education possible [B]. Special education is possible if your child cannot enter mainstream education. Do not allow anything to disturb his school and academic life. Walking does not make a person a human being, an active productive mind does.

Adolescence Provide psychological support to your child during adolescence. Try and be his friend [C]. Encourage him in the things he wants to do. Allow him to make his own decisions. Guide him gently in his relationships with his surroundings. Children need other children. Help your child make friends.

Interaction with the health care providers

Know your child well. Define your needs and express these openly to the treatment team.

Learn and perform the exercises that your physiotherapist teaches you. Ask once more if you do not understand or have difficulty performing.

Know your demands from the health care providers and from the society in general at every point in your child's life and be ready to voice them clearly. Remember that nobody can know what your child needs better than you or him [D]. Problems can only be solved if they are defined clearly and made known. The community will respond to your efforts.

Continuous care

Focus on your child's strengths, assets and interests rather than his weaknesses. People excel in the things they like doing. If your child wants to do something, support him. Praise your child frequently for his accomplishments. Do not dwell on his weaknesses. It is much easier to improve his strong points than his weaknesses. Remember, your child does not need to walk to be happy. He needs your love and attention.

Mobility is essential for the toddler who needs to explore to learn and develop skills. Sports is better than all sorts of physiotherapy for the adolescent at school.

Throughout the various therapy procedures and the anxiety of rearing a child with CP, do not miss the joy of being a father or a mother. Remember the joy of parenthood.

D	**Talking To Your Doctor**
	Here are some tips for communicating more effectively with the physician so that the visit to the doctor will be more productive.
	Make a list of your concerns. Look to the list as you talk so you will not forget anything.
	Try to arrive 10-15 minutes before the appointment time even if your doctor never runs on time.
	Have someone with you to help you listen and for emotional support if needed.
	Be conscious of the doctor's time. If you are doing well 10-15 minutes is all that is necessary.
	Use the medications as prescribed. Report effects of the medications accurately. Have a list of the medications and dosages with you all the time.
	Do not tell the doctor what to do. Work with your doctor. You are a team.
	If you do not understand what the doctor says ask him/her to repeat it.
	Do not accept ambiguous answers.
	Be impressed with physicians who send you to get a second opinion, or admit that they do not have all the answers.
	THANK the doctor for his/her time.
	Educate yourself about CP.

E	**When buying clothes look for:**
	Finger rings or other large zipper pulls
	Flat and nonskid shoes
	Raglan sleeves
	Front openings
	Short backs
	Nonrestrictive elastic
	Velcro
	Leg zippers
	Hook and loop closures
	Flat-felled seams
	Roomy
	Cut Longer in the seat to allow for sitting
	Cut shorter in the legs and arms and shorter shirts that don't get caught in wheels.
	Knit fabrics

Table modified from the website: http://geocities.com/aneecp/clothing.htm

Glossary

Apgar score A numbered score doctors use to evaluate a baby's condition at the time of birth.

apraxia Impaired ability to carry out purposeful movements in the presence of sufficient motor function.

asphyxia Lack of oxygen because of trouble with breathing or poor oxygen supply in the air.

cerebral Related to the brain.

computed tomography (CT) An imaging technique that uses X rays and a computer to create a picture of the body.

congenital Present at birth.

contracture Inability to move the joint because of stiffness in the muscles.

dysarthria Speaking difficulty because of problems in controlling the muscles needed for speech.

electroencephalogram (EEG) A method of recording electrical currents inside the brain.

electromyography A method of recording muscle and nerve activity.

failure to thrive Being behind in terms of physical growth and development.

gait analysis A method of objectively measuring walking using a camera recording, force plates, electromyography, and computer analysis.

gastrostomy A surgical procedure to create an artificial opening in the stomach.

hemianopia Defective vision or blindness in half of the field of vision of one eye.

hypertonia Increased muscle tightness.

hypotonia Decreased muscle tightness, the state of being flaccid.

hypoxic-ischemic encephalopathy Brain damage caused by poor blood and oxygen supply.

magnetic resonance imaging (MRI) An imaging technique which uses radio waves, magnetic fields, and computer analysis to create a picture of body tissues and structures.

orthoses Splints or braces used to treat problems of the muscles, ligaments, or bones of the skeletal system.

paresis or plegia Weakness or paralysis.

palsy Paralysis, inability to control voluntary movement.

reflexes Movements that the body makes automatically as a response to various stimuli.

selective dorsal root rhizotomy A surgical procedure in which selected nerves are cut to reduce spasticity in the legs.

spastic diplegia A form of cerebral palsy in which the legs are more severely affected than the arms.

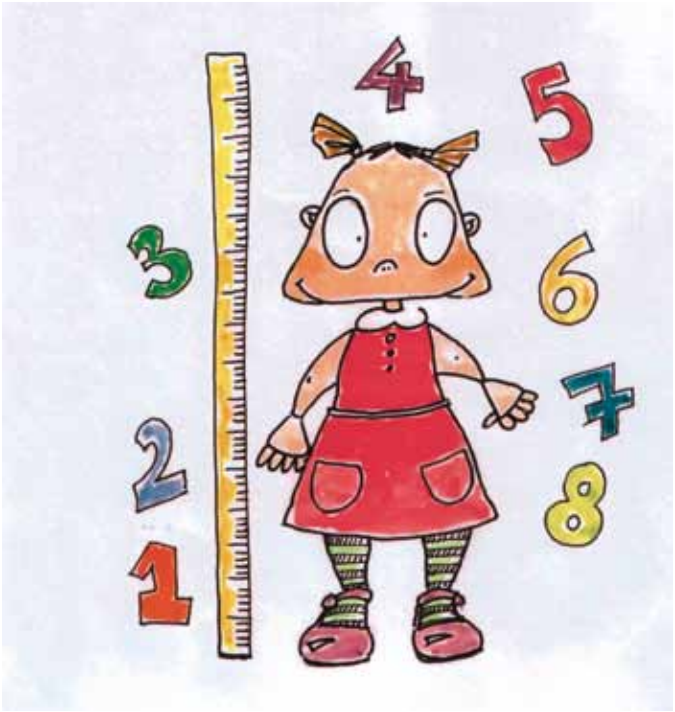
spastic hemiplegia (or hemiparesis) A form of cerebral palsy in which the arm and leg on one side of the body is affected.

spastic quadriplegia (or quadriparesis) A form of cerebral palsy in which all four limbs, the trunk and the neck are affected.

stereognosia Difficulty perceiving and identifying objects using the sense of touch.

strabismus Misalignment of the eyes.

ultrasonography A technique that uses sound waves and their echoes to form an image, called a sonogram.



This appendix covers some of the most common scales used in CP and/or information on where to find them. The Web resources that may help the readers in their further studies are also included at the very end.

Index

Developmental milestones according to Gessel and Amatruda:

Gross motor behaviour	Preambulatory skills, walking and advanced physical activities
Fine motor-adaptive behaviour	Prehension, manipulatory hand skills, application of sensorymotor skills to daily living activities
Language	Vocalization, comprehension, expression in spoken and other modes of communication
Personal-social behaviour	Acquisition of social and cultural standards of behaviour

Developmental milestones

Age	*Gross motor*	*Fine motor*	*Language*	*Personal social*
Newborn	Flexor tone, in prone turns head to side	Hands fisted, grasp reflex	Cries, turns head to sound	
2 months	Lifts head up when prone		Makes sounds, follows with eyes	Startles to loud noise, smiles responsively
4 months	Head midline, holds head when pulled to sit, in prone lifts head	Hands open, midline play, reaches for an object, grasps it and puts it to his mouth	Laughs, squeals, responds to the sounds mother makes	Recognizes bottle
6 months	Lifts head when supine, rolls from prone to supine, sits with support	Transfers objects from hand to hand	Babbles, localizes direction of sound	
7 months	Sits without suppport, rolls to prone	Bangs objects, reaches out for people	Uses single words and can say ba-ba, da-da	Differentiates between familiar person and stranger, talks to mirror image, looks for dropped things
10 months	Pulls self to sit,creeps on all fours, stands momentarily, cruises	Thumb to index grasp, pincer grasp	Uses mama and dada with meaning, shouts for attention, imitates speech	Plays peekaboo feeds himself with finger
14 months	Walks alone with arms held high, wide base, excessive knee and hip flexion, slight knee and feet valgus	Piles two cubes, scribbles spontanoeusly	Uses single words understands simple commands	Uses spoon removes clothing
18 months	Walks with lowered arms, walks backward, sits himself in chair	Emerging hand dominance, crude release	Says no, points to named body part	Imitates housework, carries and hugs dolls, drinks from cup
2 years	Begins running, jumps with both feet, walks up and down stairs	Builds eight cube tower, places pencil between thumb and fingers, draws with arm and wrist action	Two word phrases, uses verbs, follows simple directions	Puts on clothing, uses spoon well, opens door, toilet training
3 years	Runs well, pedals tricycle	Overhand throw, catches objects, copies circle	Three word sentences, future tense, asks who, what, where	Washes and dries hand and face, toilet trained, can be reasoned with
4 years	Hops on one foot, plantar arches develop	Handles a pencil like adults, draws person, cuts with scissors	Uses past tense, adjectives and adverbs, knows opposites	Cooperative play, imaginative play
5 years	Skips, tiptoes	Catches with hands, draws details	Fluent speech	Self sufficient in most ADLs

Developmental Tests

These tests describe the development of the child in various functional stages.

Denver Development Screening Test

This test evaluates the developmental deficits in infants and young children from age 1 month to 6 years in the areas of global motor function, language, fine-motor adaptation and social contact.

The Denver Developmental Screening Test (DDST) II is administered to children between birth and six years of age. It can screen children who are apparently normal for possible problems and monitor children who have high risk because of past history such as perinatal difficulties. It is not an IQ test nor will it predict what the level of the child's future intelligence and ability will be. Do not use the Denver II for diagnosis. The Denver II tests the child on twenty simple tasks and on 4 different domains. Personal - social measures the child's ability to get along with people and to take care of himself / herself. Fine Motor Adaptive test identifies the child's ability to see and to use his hands to pick up objects and to draw. Language tests determine the child's ability to hear, follow direction and to speak. Gross Motor identifies the child's ability to sit, walk and jump.

Further Reading:

Denver II Training Manual, Second Edition Revised 1992

Bayley Scales of Infant Development

This test evaluates cognition, language, social behaviour and motor functions in children from 1 to 42 months old. The purpose of the Bayley Scales of Infant Development is to diagnose developmental delay. The test takes approximately 45 minutes. The examiner gives a series of stimuli to which the child responds. The Mental Scales assess memory, learning, problem-solving ability, and verbal communication skills. The Motor Scales evaluate sitting and standing, gross motor skills and fine motor skills. The Infant Behavior Record (IBR) assesses the child's social and emotional development through a standardized description of his or her behaviour during the testing session. Scores are measured against norms for each of the 14 different age groups. The Bayley scales determine whether a child is developing normally and provide for early diagnosis and intervention in cases of developmental delay.

The Modified Ashworth scale	
0	No increase in muscle tone
1	Slight increase in tone with a catch and release or minimal resistance at end of range
2	As 1 but with minimal resistance through range following catch
3	More marked increase tone through ROM
4	Considerable increase in tone, passive movement difficult.
5	Affected part rigid

Tardieu Scale	
Velocity of stretch	
V1	As slow as possible (slower than the rate of the natural drop of the limb segment under gravity)
V2	Speed of the limb segment falling under gravity
V3	As fast as possible (faster than the rate of the natural drop of the limb segment under gravity)
Grading Tardieu scale	
Quality of muscle reaction (X)	
0	No resistance throughout the course of the passive movement
1	Slight resistance throughout the course of the passive movement
2	Clear catch at precise angle, interrupting the passive movement, followed by release
3	Fatiguable clonus (less than 10 s when maintaining the pressure) occurring at a precise angle, followed by release
4	Unfatiguable clonus (less than 10 s when maintaining the pressure) occurring at a precise angle
Angle of muscle action (V) measured relative to the position of minimal stretch of the muscle (corresponding to angle zero) for all joints except hip where it is relative to the resting anatomical position	

Gross Motor Function Classification System for Cerebral Palsy (GMFCS)

The Gross Motor Function Classification System for cerebral palsy is based on self-initiated movement with particular emphasis on sitting (truncal control) and walking. The GMFCS was developed by Robert Palisano, Peter Rosenbaum, Stephen Walter, Dianne Russell, Ellen Wood, Barbara Galuppi in the year 1997 at the CanChild Centre for Childhood Disability Research.The focus is on determining which level best represents the child's present abilities and limitations in motor function. Emphasis is on the child's usual performance in home, school, and community settings. It is therefore important to classify on ordinary performance (not best capacity), and not to include judgments about prognosis. Remember the purpose is to classify a child's present gross motor function, not to judge quality of movement or potential for improvement.

Institute for Applied Health Sciences, McMaster University
1400 Main Street West, Rm. 408, Hamilton, ON, Canada L8S 1C7 Tel: 905-525-9140 Ext. 27850 Fax: 905-522-6095
E-mail: canchild@mcmaster.ca Website: www.fhs.mcmaster.ca/canchild Dev Med Child Neurol 1997;39:214-223

1997 Palisano, R., Rosenbaum, P., Walter, S., et al 'Development and reliability of a system to classify gross motor function in children with cerebral palsy.' Developmental Medicine and Child Neurology, 39, 214-223

2008 Palisano RJ, Rosenbaum P, Bartlett D, et al 'Content Validity of the Expanded and Revised Gross Motor Function Classification System.' Developmental Medicine and Child Neurology; 50(10):744

Gross Motor Function Classification System for Cerebral Palsy (GMFCS)	
Before 2nd birthday	
Level I	Infants move in and out of sitting and floor sit with both hands free to manipulate objects. Infants crawl on hands and knees, pull to stand and take steps holding on to furniture. Infants walk between 18 months and 2 years of age without the need for any assistive mobility device.
Level II	Infants maintain floor sitting but may need to use their hands for support to maintain balance. Infants creep on their stomach or crawl on hands and knees. Infants may pull to stand and take steps holding on to furniture.
Level III	Infants maintain floor sitting when the low back is supported. Infants roll and creep forward on their stomachs.
Level IV	Infants have head control but trunk support is required for floor sitting. Infants can roll to supine and may roll to prone.
Level V	Physical impairments limit voluntary control of movement. Infants are unable to maintain antigravity head and trunk postures in prone and sitting. Infants require adult assistance to roll.

Between 2nd and 4th birthday	
Level I	Children floor sit with both hands free to manipulate objects. Movements in and out of floor sitting and standing are performed without adult assistance. Children walk as the preferred method of mobility without the need for any assistive mobility device.
Level II	Children floor sit but may have difficulty with balance when both hands are free to manipulate objects. Movements in and out of sitting are performed without adult assistance. Children pull to stand on a stable surface. Children crawl on hands and knees with a reciprocal pattern, cruise holding onto furniture and walk using an assistive mobility device as preferred methods of mobility.
Level III	Children maintain floor sitting often by "W-sitting" (sitting between flexed and internally rotated hips and knees) and may require adult assistance to assume sitting. Children creep on their stomach or crawl on hands and knees (often without reciprocal leg movements) as their primary methods of self mobility. Children may pull to stand on a stable surface and cruise short distances. Children may walk short distances indoors using an assistive mobility device and adult assistance for steering and turning.
Level IV	Children sit on a chair but need adaptive seating for trunk control and to maximize hand function. Children move in and out of chair sitting with assistance from an adult or a stable surface to push or pull up on with their arms. Children may at best walk short distances with a walker and adult supervision but have difficulty turning and maintaining balance on uneven surfaces. Children are transported in the community. Children may achieve self-mobility using a power wheelchair.
Level V	Physical impairments restrict voluntary control of movement and the ability to maintain antigravity head and trunk postures. All areas of motor function are limited. Functional limitations in sitting and standing are not fully compensated for through the use of adaptive equipment and assistive technology. At Level V, children have no means of independent mobility and are transported. Some children achieve self-mobility using a power wheelchair with extensive adaptations.

Between 4th and 6th birthday	
Level I	Children get into and out of, and sit in, a chair without the need for hand support. Children move from the floor and from chair sitting to standing without the need for objects for support. Children walk indoors and outdoors, and climb stairs. Emerging ability to run and jump.
Level II	Children sit in a chair with both hands free to manipulate objects. Children move from the floor to standing and from chair sitting to standing but often require a stable surface to push or pull up on with their arms. Children walk without the need for any assistive mobility device indoors and for short distances on level surfaces outdoors. Children climb stairs holding onto a railing but are unable to run or jump.
Level III	Children sit on a regular chair but may require pelvic or trunk support to maximize hand function. Children move in and out of chair sitting using a stable surface to push on or pull up with their arms. Children walk with an assistive mobility device on level surfaces and climb stairs with assistance from an adult. Children frequently are transported when travelling for long distances or outdoors on uneven terrain.
Level IV	Children sit on a chair but need adaptive seating for trunk control and to maximize hand function. Children move in and out of chair sitting with assistance from an adult or a stable surface to push or pull up on with their arms. Children may at best walk short distances with a walker and adult supervision but have difficulty turning and maintaining balance on uneven surfaces. Children are transported in the community. Children may achieve self-mobility using a power wheelchair.

Level V	Physical impairments restrict voluntary control of movement and the ability to maintain antigravity head and trunk postures. All areas of motor function are limited. Functional limitations in sitting and standing are not fully compensated for through the use of adaptive equipment and assistive technology. At Level V, children have no means of independent mobility and are transported. Some children achieve self-mobility using a power wheelchair with extensive adaptations.

Between 6th and 12th birthday	
Level I	Children walk indoors and outdoors, and climb stairs without limitations. Children perform gross motor skills including running and jumping but speed, balance, and coordination are reduced.
Level II	Children walk indoors and outdoors, and climb stairs holding onto a railing but experience limitations walking on uneven surfaces and inclines, and walking in crowds or confined spaces. Children have at best only minimal ability to perform gross motor skills such as running and jumping.
Level III	Children walk indoors or outdoors on a level surface with an assistive mobility device. Children may climb stairs holding onto a railing. Depending on upper limb function, children propel a wheelchair manually or are transported when travelling for long distances or outdoors on uneven terrain.
Level IV	Children may maintain levels of function achieved before age 6 or rely more on wheeled mobility at home, school, and in the community. Children may achieve self-mobility using a power wheelchair.
Level V	Physical impairments restrict voluntary control of movement and the ability to maintain antigravity head and trunk postures. All areas of motor function are limited. Functional limitations in sitting and standing are not fully compensated for through the use of adaptive equipment and assistive technology. At level V, children have no means of independent mobility and are transported. Some children achieve self-mobility using a power wheelchair with extensive adaptations.

The Quality of Upper Extremity Skills Test - QUEST

The QUEST is a measure designed to evaluate movement patterns and hand function in children with cerebral palsy. Validation studies have been conducted with children aged 18 months to 8 years. To evaluate quality of upper extremity function in four domains: dissociated movement, grasp, protective extension, and weight bearing. It evaluates quality of movement in children with cerebral palsy. It is administered within a play context. Items are related to quality of movement, not to chronological age. There are 36 items assessing dissociated movements, grasp, protective extension, and weight bearing. 30 - 45 minutes. Validation studies have been completed with children with cerebral palsy.
DeMatteo, C., Law, M., Russell, D., Pollock, N., Rosenbaum, P., & Walter, S. (1992). QUEST: Quality of Upper Extremity Skills Test. Hamilton, ON: McMaster University, Neurodevelopmental Clinical Research Unit
DeMatteo, C., Law, M., Russell, D., Pollock, N., Rosenbaum, P., & Walter, S. (1993). The reliability and validity of Quality of Upper Extremity Skills Test. Physical and Occupational Therapy in Pediatrics 13(2), 1-18.

Canadian Occupational Performance Measure (COPM)

The Canadian Occupational Performance Measure (COPM) is a measurement tool that assists therapists in using a family-centred approach to service delivery by indicating the family's priorities. It assists therapists in using a client-centred approach to service delivery by indicating the family's priorities. It thus enables therapy to be individualized and targeted to the areas of greatest need and offers an effective system of measuring the outcomes of therapy. This measure is available from the Canadian Association of Occupational Therapists (CAOT). www.caot.ca

The Pediatric Evaluation of Disability Inventory (PEDI)

The PEDI is developed to measure functional status and functional change in self care activities, mobility activities and social function. Self care consists of feeding, grooming, dressing and toileting, mobility consists of car, chair, tub and toilet transfers, indoor, outdoor walking and stairs; social function consists of comprehension, speech, interactions with friends and in the community. Capability is measured by the identification of functional skills for which the child has demonstrated mastery and competence. Functional performance is measured by the level of caregiver assistance needed to accomplish major functional activities such as eating or outdoor locomotion. A modifications scale provides a measure of environmental modifications and equipment used by the child in routine daily activities.

The PEDI compares the child' scores to an age matched normal group of children, or the child's performance with a total possible score of 100 which corresponds to the maximum score a normal 7 year old can get. It is useful both for the diagnosis of functional delay and also for assessing progress in therapy. The PEDI was designed primarily for children from 6 months of age to 7 years, however, it can also be used for the evaluation of older children if their functional abilities fall below that expected of seven-year-old children without disabilities. Scores are recorded in a booklet which also contains a summary score sheet that can be used to construct a profile of the child's performance across the different domains and scales. A software program for data entry, scoring, and generation of individual summary profiles is also available for IBM-compatible computers.

The PEDI can be administered by clinicians and familiar with the child, or by interview of the parent. The amount of time required for the parent interview is about 45 minutes. Administration guidelines, criteria for scoring each item, and examples are given in the manual. The manual also contains information on instrument development and validation, including normative information as well as data from several clinical samples.

The PEDI can be ordered from: Center for Rehabilitation Effectiveness, Sargent College of Health and Rehabilitation Sciences, Boston University, Boston, MA 02215 Phone: 617-358-0175 Fax: 617-388-1355 email: pandres@bu.edu website: www.bu.edu/cre/pedi

GROSS MOTOR FUNCTION MEASURE (GMFM)
SCORE SHEET (GMFM-88 and GMFM-66 scoring)
Version 1.0

Child's Name: _____ ID #: _____
Assessment date: _____ GMFCS Level: ☐ ☐ ☐ ☐ ☐
year / month/ day I II III IV V
Date of birth: _____
year / month/ day
Chronological age: _____
month/ day Testing conditions (eg. room, clothing, time, others present):

Evaluator's Name:

The GMFM is a standardized observational instrument designed and validated to measure change in gross motor function over time in children with cerebral palsy. The scoring kay is meant to be a general guideline. However, most of the items have specific dascriptors for each score. It is imperative that the guidelines contained in the manual be used for scoring each item.

SCORING KEY 0= does not initiate
1= initiates
2= partially completes
3= completes
NT= Not tested [used for the GAME scoring]

It is now importand to differentiate a true score of "0" (child does not initiate) from an item which is Not Tested (NT) if you are interested in using the GMFM-66 Ability Estimator Software.

* The GMFM-66 Gross Motor Ability Estimator (GMAE) software is available with the GMFM manual (2002). The advantage of the software is the conversion of the ordinal scale into an interval scale. This will allow for a more accurate estimate of the child's ability and provide a measure that is equally responsive to change a cross the spectrum of ability levels. items that are used in the calculation of the GMFM-66 score are shaded and identified with a asterisk (*). The GMFM-66 is only valid for use with children who have CP.

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Website:www.fhs.mcmaster.ca/canchild

GMFCS level is a rating of severity of motor function. Definitions are found in Appendix I of the GMFM manual (2002).

Check (✓) the appropriate score: if an item is not tested (NT), circle the item number in the left column.

Item	A: LYING & ROLLING	SCORE
1.	SUP, HEAD IN MIDLINE: turns head with extremities symmetrical	0☐ 1☐ 2☐ 3☐
* 2.	SUP: brings hands to midline, fingers one with the other	0☐ 1☐ 2☐ 3☐
3.	SUP: lifts head 45°	0☐ 1☐ 2☐ 3☐
4.	SUP: flexes r hip and knee through full range	0☐ 1☐ 2☐ 3☐
5.	SUP: flexes l hip and knee through full range	0☐ 1☐ 2☐ 3☐
* 6.	SUP: reaches out with r arm, hand crosses midline toward toy	0☐ 1☐ 2☐ 3☐
* 7.	SUP: reaches out with l arm, hand crosses midline toward toy	0☐ 1☐ 2☐ 3☐
8.	SUP: rolls to pr over r side	0☐ 1☐ 2☐ 3☐
9.	SUP: rolls to pr over l side	0☐ 1☐ 2☐ 3☐
*10.	PR: lifts head up right	0☐ 1☐ 2☐ 3☐
11.	PR ON FOREARMS: lifts head upright, elbows ext, chest raised	0☐ 1☐ 2☐ 3☐
12.	PR ON FOREARMS: weight on r forearm, fully extends opposite arm forward	0☐ 1☐ 2☐ 3☐
13.	PR ON FOREARMS: weight on l forearm, fully extends opposite arm forward	0☐ 1☐ 2☐ 3☐
14.	PR: rolls to sup over r side	0☐ 1☐ 2☐ 3☐
15.	PR: rolls to sup over l side	0☐ 1☐ 2☐ 3☐
16.	PR: pivots to r 90° using extremities	0☐ 1☐ 2☐ 3☐
17.	PR: pivots to l 90° using extremities	0☐ 1☐ 2☐ 3☐
	TOTAL DIMENSION A	☐

Check (✓) the appropriate score: if an item is not tested (NT), circle the item number in the left column.

Item B: SITTING	SCORE
*18. SUP: HANDS GRASPED BY EXAMINER: pulls self to sitting with head control _____	0☐ 1☐ 2☐ 3☐
19. SUP: rolls to r side, attains sitting	0☐ 1☐ 2☐ 3☐
20. SUP: rolls to l side, attains sitting	0☐ 1☐ 2☐ 3☐
*21. SIT ON MAT, SUPPORTED AT THORAX BY THERAPIST: lifts head upright, maintain 3 seconds	0☐ 1☐ 2☐ 3☐
*22. SIT ON MAT, SUPPORTED AT THORAX BY THERAPIST: lifts head midline, maintains 10 seconds	0☐ 1☐ 2☐ 3☐
*23. SIT ON MAT, ARM(S) PROPPING: maintains 5 seconds	0☐ 1☐ 2☐ 3☐
*24. SIT ON MAT: maintains, arms free, 3 seconds	0☐ 1☐ 2☐ 3☐
*25. SIT ON MAT WITH SMALL TOY IN FRONT:leans forward,touches toy, re-erects without arm propping	0☐ 1☐ 2☐ 3☐
*26. SIT ON MAT: touches toy placed 45° behind child's r side, returns to start	0☐ 1☐ 2☐ 3☐
*27. SIT ON MAT: touches toy placed 45° behind child's l side, returns to start	0☐ 1☐ 2☐ 3☐
28. R SIDE SIT: maintains, arms free, 5 seconds	0☐ 1☐ 2☐ 3☐
29. L SIDE SIT: maintains, arms free, 5 seconds	0☐ 1☐ 2☐ 3☐
*30. SIT ON MAT: lowers to pr with control	0☐ 1☐ 2☐ 3☐
*31. SIT ON MAT WITH FEET IN FRONT: attains 4 points over r side	0☐ 1☐ 2☐ 3☐
*32. SIT ON MAT WITH FEET IN FRONT: attains 4 point over l side	0☐ 1☐ 2☐ 3☐
33. SIT ON MAT: pivots 90°, without arms assisting	0☐ 1☐ 2☐ 3☐
*34. SIT ON BENCH: maintains, arms and free,10 seconds	0☐ 1☐ 2☐ 3☐
*35. STD: attains sit on small bench	0☐ 1☐ 2☐ 3☐
*36. ON THE FLOOR: Attains sit on small bench	0☐ 1☐ 2☐ 3☐
*37. ON THE FLOOR: attains sit on large bench	0☐ 1☐ 2☐ 3☐

TOTAL DIMENSION B ☐

Check (✓) the appropriate score: if an item is not tested (NT), circle the item number in the left column.

Item	C: CRAWLING & KNEELING	SCORE
38.	PR: creeps forward 1.8 M (6')	0☐ 1☐ 2☐ 3☐
*39.	4 POINT: maintains, weight on hands and knees, 10 seconds	0☐ 1☐ 2☐ 3☐
*40.	4 POINT: attains sit arms free	0☐ 1☐ 2☐ 3☐
*41.	PR: attains 4 point,weight on hands and knees	0☐ 1☐ 2☐ 3☐
*42.	4 POINT: reaches forward with r arm, hand above shoulder level	0☐ 1☐ 2☐ 3☐
*43.	4 POINT: reaches forward with l arm, hand above shoulder level	0☐ 1☐ 2☐ 3☐
*44.	4 POINT: crawls or hitches forward 1.8 M (6')	0☐ 1☐ 2☐ 3☐
*45.	4 POINT: crawls reciprocally forward 1.8 M (6')	0☐ 1☐ 2☐ 3☐
*46.	4 POINT: crawls up 4 steps on hands and knees/feet	0☐ 1☐ 2☐ 3☐
47.	4 POINT: crawls backwards down 4 steps on hands and knees/feet	0☐ 1☐ 2☐ 3☐
*48.	SIT ON MAT: attains high kn using arms, maintains, arms free,10 seconds	0☐ 1☐ 2☐ 3☐
49.	HIGH KN: attains half kn on r knee using arms, maintains,arm free,10 seconds	0☐ 1☐ 2☐ 3☐
50.	HIGH KN: attains half kn on l knee using arms, maintains,arm free,10 seconds	0☐ 1☐ 2☐ 3☐
*51.	HIGH KN: kn walks forward 10 step, arms free	0☐ 1☐ 2☐ 3☐

TOTAL DIMENSION C ☐

Check (✓) the appropriate score: if an item is not tested (NT), circle the item number in the left column.

Item D: STANDING	SCORE
***52. ON THE FLOOR: pulls to std at large bench**	0☐ 1☐ 2☐ 3☐
*53. STD: maintains, arm free, 3 seconds	0☐ 1☐ 2☐ 3☐
*54. STD: holding on to large bench with one hand, lifts r foot, 3 seconds	0☐ 1☐ 2☐ 3☐
*55. STD: holding on to large bench with one hand, lifts l foot, 3 seconds	0☐ 1☐ 2☐ 3☐
*56. STD: maintains, arms free, 20 seconds	0☐ 1☐ 2☐ 3☐
*57. STD: lifts l foot, arms free, 10 seconds	0☐ 1☐ 2☐ 3☐
*58. STD: lifts r foot, arms free, 10 seconds	0☐ 1☐ 2☐ 3☐
*59. SIT ON SMALL BENCH: attains std without using arms	0☐ 1☐ 2☐ 3☐
*60. HIGH KN: attains std trough half kn on r knee,without using arms	0☐ 1☐ 2☐ 3☐
*61. HIGH KN: attains std trough half kn on l knee,without using arms	0☐ 1☐ 2☐ 3☐
*62. STD: lowers to sit on floor with control, arms free	0☐ 1☐ 2☐ 3☐
*63. STD: attains squat, arms free	0☐ 1☐ 2☐ 3☐
*64. STD: picks up object from floor, arms free, returns to stand	0☐ 1☐ 2☐ 3☐
TOTAL DIMENSION D	☐

Check (✓) the appropriate score: if an item is not tested (NT), circle the item number in the right column

Item E: WALKING, RUNNING & JUMPING	**SCORE**
*65. STD, 2 HANDS ON LARGE BENCH: cruises 5 steps to r	0☐ 1☐ 2☐ 3☐
*66. STD, 2 HANDS ON LARGE BENCH: cruises 5 steps to l	0☐ 1☐ 2☐ 3☐
*67. STD, 2 HANDS HELD: walks forward 10 steps	0☐ 1☐ 2☐ 3☐
*68. STD, 1 HANDS HELD: walks forward 10 steps	0☐ 1☐ 2☐ 3☐
*69. STD: walks forward 10 steps	0☐ 1☐ 2☐ 3☐
*70. STD: walks forward 10 steps,stops,turns 180°,returns	0☐ 1☐ 2☐ 3☐
*71. STD: walks forward 10 steps	0☐ 1☐ 2☐ 3☐
*72. STD: walks forward 10 steps,carrying a large object with 2 hands	0☐ 1☐ 2☐ 3☐
*73. STD: walks forward 10 consecutive steps between parallel lines 20 cm (8”) apart	0☐ 1☐ 2☐ 3☐
*74. STD: walks forward 10 consecutive steps on a straight line 2 cm (3/4”) wide	0☐ 1☐ 2☐ 3☐
*75. STD: steps over stick at knee level, r foot leading	0☐ 1☐ 2☐ 3☐
*76. STD: steps over stick at knee level, l foot leading	0☐ 1☐ 2☐ 3☐
*77. STD: runs 4.5 M (15’), stops & returns	0☐ 1☐ 2☐ 3☐
*78. STD: kicks ball with r foot	0☐ 1☐ 2☐ 3☐
*79. STD: kicks ball with l foot	0☐ 1☐ 2☐ 3☐
*80. STD: jumps 30 cm (12”) high, both feet simultaneously	0☐ 1☐ 2☐ 3☐
*81. STD: jumps forward 30 cm (12”), both feet simultaneously	0☐ 1☐ 2☐ 3☐
*82. STD ON R FOOT: hops on r foot 10 times within a 60 cm (24”) circle	0☐ 1☐ 2☐ 3☐
*83. STD ON L FOOT: hops on l foot 10 times within a 60 cm (24”) circle	0☐ 1☐ 2☐ 3☐
*84. STD,HOLDING 1 RAIL: walks up 4 steps, holding 1 rail,alternating feet	0☐ 1☐ 2☐ 3☐
*85. STD,HOLDING 1 RAIL: walks down 4 steps, holding 1 rail,alternating feet	0☐ 1☐ 2☐ 3☐
*86. STD: walks up 4 steps, alternating feet	0☐ 1☐ 2☐ 3☐
*87. STD: walks down 4 steps, alternating feet	0☐ 1☐ 2☐ 3☐
*88. STD ON 15 cm (6”) STEP: jumps off, both feet simultaneously	0☐ 1☐ 2☐ 3☐
TOTAL DIMENSION E	☐

Was this assessment indicative of this child’s “regular” performance? YES ☐ NO ☐

COMMENTS:

GMFM RAW SUMMARY SCORE

DIMENSION	CALCULATION OF DIMENSION % SCORES	GOAL AREA
A. Lying & Rolling	$\frac{\text{Total Dimension A}}{51} = \frac{\quad}{51} \times 100 = \quad\%$	A ☐
B. Sitting	$\frac{\text{Total Dimension B}}{60} = \frac{\quad}{60} \times 100 = \quad\%$	B ☐
C. Crawling & Kneeling	$\frac{\text{Total Dimension C}}{42} = \frac{\quad}{42} \times 100 = \quad\%$	C ☐
D. Standing	$\frac{\text{Total Dimension D}}{39} = \frac{\quad}{39} \times 100 = \quad\%$	D ☐
E. Walking, Running & Jumping	$\frac{\text{Total Dimension E}}{72} = \frac{\quad}{72} \times 100 = \quad\%$	E ☐

$$\textit{TOTAL SCORE} = \frac{\%A + \%B + \%C + \%D + \%E}{\text{Total \# of Dimensions}}$$

$$= \frac{\quad + \quad + \quad + \quad + \quad}{5} = \frac{\quad}{5} = \quad\%$$

GOAL TOTAL SCORE = $\frac{\text{Sum of \% scores foe each dimension identified as a goal area}}{\text{\# of Goal areas}}$

$$= \frac{\quad}{\quad} = \quad\%$$

GMFM-66 Gross Motor Ability Estimator Score [1]

GMFM-66 Score = _____ _____ to _____ 95% Confidence Intervals

previous GMFM-66 Score = _____ _____ to _____ 95% Confidence Intervals

change in GMFM-66 = _____

[1] from the Gross Motor Ability Estimator (GMAE) Software

TESTING WITH AIDS/ORTHOSES

Indicate below with a check (✓) which aid/orthosis was used and what dimension it was first applied. (There may be more than one).

AID		DIMENSION	ORTHOSIS		DIMENSION
Rollator/Pusher	☐	_____	Hip Control	☐	_____
Walker	☐	_____	Knee Control	☐	_____
H Frame Crutches	☐	_____	Ankle-Foot Control	☐	_____
Crutches	☐	_____	Foot Control	☐	_____
Quad Cane	☐	_____	Shoes	☐	_____
Cane	☐	_____	None	☐	_____
None	☐	_____	Other	☐	_____
Other	☐	_____			(please specify)
		(please specify)			

RAW SUMMARY SCORE USING AIDS/ORTHOSES

DIMENSION	CALCULATION OF DIMENSION % SCORES	GOAL AREA
F. Lying & Rolling	Total Dimension A / 51 = / 51 x 100= %	A
G. Sitting	Total Dimension B / 60 = / 60 x 100= %	B
H. Crawling & Kneeling	Total Dimension C / 42 = / 42 x100= %	C
I. Standing	Total Dimension D / 39 = / 39 x100= %	D
J. Walking, Running & Jumping	Total Dimension E / 72 = / 72 x 100= %	E

TOTAL SCORE = (%A + %B + %C + %D + %E) / Total # of Dimensions

= (+ + + +) / 5 = / 5 = %

GOAL TOTAL SCORE = Sum of % scores foe each dimension identified as a goal area / # of Goal areas

= = %

GMFM-66 Gross Motor Ability Estimator Score [1]

GMFM-66 Score = to
95% Confidence Intervals

previous GMFM-66 Score = to
95% Confidence Intervals

change in GMFM-66 =

[1] from the Gross Motor Ability Estimator (GMAE) Software

WEE Functional Independence Measure (WeeFIM) ®

The WeeFIM is an adaptation of the Functional Independence Measure for adults. It was developed to measure the need for assistance and the severity of disability in children between the ages of 6 months and 7 years. It may be used with children above the age of 7 years as long as their functional abilities are below those expected of children aged 7 who do not have disabilities. It measures level of independence in self-care, sphincter control, mobility, locomotion, communication and social function. It is a data set of 18 items that measure functional performance in 3 domains:

The WeeFIM		
Self-Care	*Mobility*	*Cognitive*
Eating	Transfers: Chair/Wheelchair	Comprehension
Grooming	Transfers: Toilet	Expression
Bathing	Transfers: Tub/Shower	Social Interaction
Dressing: Upper Body	Locomotion: Walk/Wheelchair	Problem Solving
Dressing: Lower Body	Stairs	Memory
Toileting		
Bladder Management		
Bowel Management		

Web Resources

www.aacpdm.org
This organization is a multidisciplinary scientific society devoted to the study of cerebral palsy and other childhood onset disabilities, to promoting professional education for the treatment and management of these conditions, and to improving the quality of life for people with these disabilities.

www.ucpa.org
This organization tries to ensure the inclusion of persons with disabilities in every facet of society. The UCP's mission is to advance the independence, productivity and full citizenship of people with cerebral palsy and other disabilities, through commitment to the principles of independence, inclusion and self-determination.

www.wemove.org
This website is a comprehensive resource for movement disorder information and movement disorder activities on the web.

www.mdvu.org
The Movement Disorder Virtual University is the healthcare professional's source for movement disorder news, resources and educational activities. Explore the MDVU virtual campus to find the latest information on emerging clinical advances and therapeutic approaches, interactive learning modules (with CME credit), case studies, practice tools, teaching materials and opportunities for peer interaction.

www.ispoint.org
This is the website of the The International Society for Prosthetics and Orthotics (ISPO)

www.nlm.nih.gov/medlineplus/cerebralpalsy.html
The MedlinePlus is a source of good health information from the world's largest medical library, the National Library of Medicine. Health professionals and consumers alike can find information that is authoritative and up to date.

www.fhs.mcmaster.ca/canchild/
CanChild is a centre for childhood disability research that seeks to maximize the life quality of children and youth with disabilities and their families.

www.ninds.nih.gov/health_and_medical/disorders/cerebral_palsy.htm
National Institute of Neurological Disorders and Stroke is dedicated to support biomedical research on disorders of the brain and nervous system. The website provides information about cerebral palsy and the latest research being done on the subject.

www.familyvoices.org
This website aims to achieve family-centered care for all children and youth with special health care needs and/or disabilities.

www.scope.org.uk
The Scope is a disability organisation in England and Wales whose focus is people with cerebral palsy. Its aim is that disabled people achieve equality in a society in which they are as valued and have the same human and civil rights as everyone else.

www.pediatricapta.org/index.cfm
This website is the pediatrics section of the American Association of Physical Therapists. It contains comprehensive information on pediatric physical therapy.

www.kidsource.com/NICHCY/cerebral_palsy.html
This is part of the website of National Information Center for Children and Youth with Disabilities (NICHY) It contains general information about cerebral palsy.

www.bobath.co.uk
The Bobath Center's website provides information on CP as well as the Bobath method of treatment.

www.modimes.org
The March of Dimes is a foundation to help children with birth defects. This website provides information on CP.

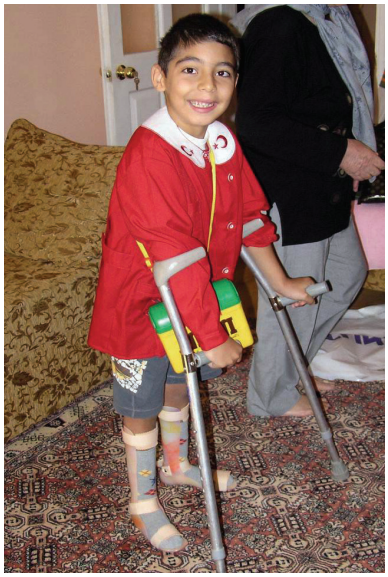
www.conductive-education.org.uk
This site provides extensive information on conductive education.

www.udsmr.org
This site provides information about the Wee functional independence measure.

Cerebral Palsy is one of the devastating disorders in medicine. This relatively common problem occurs in babies, lasts for a lifetime and creates disability ranging from mild to severe. The lesion cannot be cured but the consequences can be minimized with proper treatment.

The past decades have witnessed a great deal of change and improvement in the management of CP. Procedures have been modified to be more effective and at the same time minimize the impact of therapy, bracing and surgery on the child's life. However, this changing and increasing knowledge did not reach many parts of the world.

This book aims to provide concise, up-to-date information on the basic aspects and new approaches to treatment of musculoskeletal problems of children with CP. We hope that it will be useful in the correction of problems that cause lifelong disability for millions of children worldwide.



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