Cerebral Palsy

Hope Through Research

National Institute of Neurological Disorders
and Stroke
National Institutes of Health

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What is cerebral palsy?

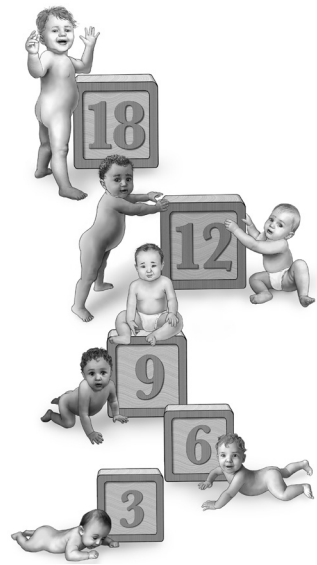
Cerebral palsy refers to a group of neurological disorders that appear in infancy or early childhood and permanently affect body movement and muscle coordination. Cerebral palsy (CP) is caused by damage to or abnormalities inside the developing brain that disrupt the brain's ability to control movement and maintain posture and balance. The term *cerebral** refers to the brain; *palsy* refers to the loss or impairment of motor function.

Cerebral palsy affects the motor area of the brain's outer layer (called the cerebral cortex), the part of the brain that directs muscle movement.

In some cases, the cerebral motor cortex hasn't developed normally during fetal growth. In others, the damage is a result of injury to the brain either before, during, or after birth. In either case, the damage is not repairable and the disabilities that result are permanent.

Children with CP exhibit a wide variety of symptoms, including:

- lack of muscle coordination when performing voluntary movements (*ataxia*);
- stiff or tight muscles and exaggerated reflexes (*spasticity*);



Infants with cerebral palsy often have developmental delay, in which they are slow to reach normal developmental milestones, shown above, such as learning to sit, crawl, or walk.

*Terms in italics are defined in the Glossary, page 33.

- weakness in one or more arm or leg;
- walking on the toes, a crouched gait, or a "scissored" gait;
- variations in muscle tone, either too stiff or too floppy;
- excessive drooling or difficulties swallowing or speaking;
- shaking (*tremor*) or random involuntary movements;
- delays in reaching motor skill milestones; and
- difficulty with precise movements such as writing or buttoning a shirt.

The symptoms of CP differ in type and severity from one person to the next, and may even change in an individual over time. Symptoms may vary greatly among individuals, depending on which parts of the brain have been injured. All people with cerebral palsy have problems with movement and posture, and some also have some level of intellectual disability, seizures, and abnormal physical sensations or perceptions, as well as other medical disorders. People with CP also may have impaired vision or hearing, and language, and speech problems.

CP is the leading cause of childhood disabilities, but it doesn't always cause profound disabilities. While one child with severe CP might be unable to walk and need extensive, lifelong care, another child with mild CP might be only slightly awkward

and require no special assistance. The disorder isn't progressive, meaning it doesn't get worse over time. However, as the child gets older, certain symptoms may become more or less evident.

A study by the Centers for Disease Control and Prevention shows the average prevalence of cerebral palsy is 3.3 children per 1,000 live births.

There is no cure for cerebral palsy, but supportive treatments, medications, and surgery can help many individuals improve their motor skills and ability to communicate with the world.

What are the early signs?

The signs of cerebral palsy usually appear in the early months of life, although specific diagnosis may be delayed until age two years or later. Infants with CP frequently have *developmental delay*, in which they are slow to reach developmental milestones such as learning to roll over, sit, crawl, or walk. Some infants with CP have abnormal muscle tone. Decreased muscle tone (*hypotonia*) can make them appear relaxed, even floppy. Increased muscle tone (*hypertonia)* can make them seem stiff or rigid. In some cases, an early period of hypotonia will progress to hypertonia after the first 2 to 3 months of life. Children with CP may also have unusual posture or favor one side of the body when they reach, crawl, or move. It is important to note that some children without CP also might have some of these signs.

Some Early Warning Signs of CP:

In a Baby Younger Than 6 Months of Age

- His head lags when you pick him up while he's lying on his back
- He feels stiff
- He feels floppy
- When held cradled in your arms, he seems to overextend his back and neck, constantly acting as if he is pushing away from you.
- When you pick him up, his legs get stiff and they cross or scissor

In a Baby Older Than 6 Months of Age

- She doesn't roll over in either direction
- She cannot bring her hands together
- She has difficulty bringing her hands to her mouth
- She reaches out with only one hand while keeping the other fisted

In a Baby Older Than 10 Months of Age

- He crawls in a lopsided manner, pushing off with one hand and leg while dragging the opposite hand and leg
- He scoots around on his buttocks or hops on his knees, but doesn't crawl on all fours

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Centers for Disease Control and Prevention, June 2013

What causes cerebral palsy?

Cerebral palsy is caused by abnormal development of part of the brain or by damage to parts of the brain that control movement. This damage can occur before, during, or shortly after birth. The majority of children have *congenital cerebral palsy* (that is, they were born with it), although it may not be detected until months or years later. A small number of children have *acquired cerebral palsy*, which means the disorder begins after birth. Some causes of acquired cerebral palsy include brain damage in the first few months or years of life, brain infections such as bacterial meningitis or viral encephalitis, problems with blood flow to the brain, or head injury from a motor vehicle accident, a fall, or child abuse.

In many cases, the cause of cerebral palsy is unknown. Possible causes include genetic abnormalities, congenital brain malformations, maternal infections or fevers, or fetal injury, for example. The following types of brain damage may cause its characteristic symptoms:

Damage to the white matter of the brain (*periventricular leukomalacia*, or PVL). The white matter of the brain is responsible for transmitting signals inside the brain and to the rest of the body. Damage from PVL looks like tiny holes in the white matter of an infant's brain. These gaps in brain tissue interfere with the normal transmission of signals. Researchers have identified a period of selective vulnerability in the developing fetal brain, a period of time between 26 and 34 weeks of *gestation*, in which periventricular white matter is particularly sensitive to insults and injury.

Abnormal development of the brain (*cerebral dysgenesis*). Any interruption of the normal process of brain growth during fetal development can cause brain malformations that interfere with the transmission of brain signals. Mutations in the genes that control brain development during this early period can keep the brain from developing normally. Infections, fevers, trauma, or other conditions that cause unhealthy conditions in the womb also put an unborn baby's nervous system at risk.

Bleeding in the brain (intracranial hemorrhage)**.** Bleeding inside the brain from blocked or broken blood vessels is commonly caused by fetal stroke. Some babies suffer a stroke while still in the womb because of blood clots in the *placenta* that block blood flow in the brain. Other types of fetal stroke are caused by malformed or weak blood vessels in the brain or by blood-clotting abnormalities. Maternal high blood pressure (hypertension) is a common medical disorder during pregnancy and is more common in babies with fetal stroke. Maternal infection, especially pelvic inflammatory disease, has also been shown to increase the risk of fetal stroke.

Bleeding in the brain (*asphyxia*). Asphyxia, a lack of oxygen in the brain caused by an interruption in breathing or poor oxygen supply, is common for a brief period of time in babies due to the stress of labor and delivery. If the supply of oxygen is cut off or reduced for lengthy periods, an infant can develop a type of brain damage called *hypoxic-ischemic encephalopathy*, which destroys tissue in the cerebral motor cortex and other areas of the brain. This kind of damage can also be caused

by severe maternal low blood pressure, rupture of the uterus, detachment of the placenta, or problems involving the umbilical cord, or severe trauma to the head during labor and delivery.

The risk of cerebral palsy is higher among low birthweight babies and those born prematurely.

What are the risk factors?

There are some medical conditions or events that can happen during pregnancy and delivery that may increase a baby's risk of being born with cerebral palsy. These risks include:

Blood type incompatibility between mother and child. *Rh incompatibility* is a condition that develops when a mother's Rh blood type (either positive or negative) is different from the blood type of her baby. The mother's system doesn't tolerate the baby's different blood type and her body will begin to make antibodies that will attack and kill her baby's blood cells, which can cause brain damage.

Exposure to toxic substances. Mothers who have been exposed to toxic substances during pregnancy, such as methyl mercury, are at a heightened risk of having a baby with cerebral palsy.

Infections during pregnancy. Infections such as toxoplasmosis, rubella (German measles), cytomegalovirus, and herpes can infect the womb and placenta. Inflammation triggered by infection

may then go on to damage the developing nervous system in an unborn baby. Maternal fever during pregnancy or delivery can also set off this kind of inflammatory response.

Low birthweight and premature birth. Premature babies (born less than 37 weeks into pregnancy) and babies weighing less than 5 ½ pounds at birth have a much higher risk of developing cerebral palsy than full-term, heavier weight babies. Tiny babies born at very early gestational ages are especially at risk.

Mothers with thyroid abnormalities, intellectual disability, excess protein in the urine, or seizures. Mothers with any of these conditions are slightly more likely to have a child with CP.

Multiple births. Twins, triplets, and other multiple births — even those born at term — are linked to an increased risk of cerebral palsy. The death of a baby's twin or triplet further increases the risk.

There are also medical conditions during labor and delivery, and immediately after delivery, that act as warning signs for an increased risk of CP. However, most of these children will not develop CP. Warning signs include:

Breech presentation. Babies with cerebral palsy are more likely to be in a breech position (feet first) instead of head first at the beginning of labor. Babies who are unusually floppy as fetuses are more likely to be born in the breech position.

Complicated labor and delivery. A baby who has vascular or respiratory problems during labor and delivery may already have suffered brain damage or abnormalities.

Jaundice. More than 50 percent of newborns develop jaundice (a yellowing of the skin or whites of the eyes) after birth when *bilirubin*, a substance normally found in bile, builds up faster than their livers can break it down and pass it from the body. Severe, untreated jaundice can kill brain cells and can cause deafness and CP.

Low Apgar score. The Apgar score is a numbered rating that reflects a newborn's physical health. Doctors periodically score a baby's heart rate, breathing, muscle tone, reflexes, and skin color during the first minutes after birth. A low score at 10-20 minutes after delivery is often considered an important sign of potential problems such as CP.

Seizures. An infant who has seizures faces a higher risk of being diagnosed later in childhood with CP.

Small for gestational age. Babies born smaller than normal for their gestational age are at risk for cerebral palsy because of factors that kept them from growing naturally in the womb.

Can cerebral palsy be prevented?

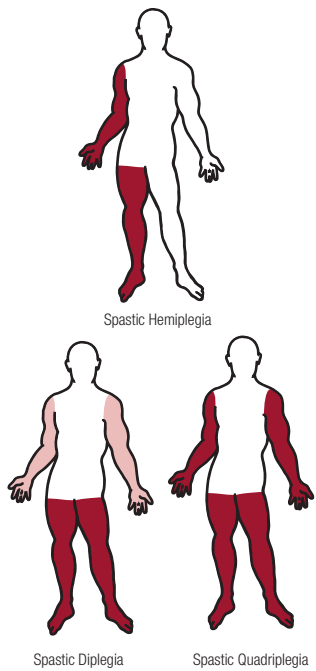
Cerebral palsy related to genetic abnormalities cannot be prevented, but a few of the risk factors for congenital cerebral palsy can be managed or avoided. For example, *rubella*, or German measles, is preventable if women are vaccinated against the disease before becoming pregnant. Rh incompatibilities can also be managed early in pregnancy. Acquired cerebral palsy, often due to head injury, is often preventable using common safety tactics, such as using car seats for infants and toddlers.

What are the different forms?

The specific forms of cerebral palsy are determined by the extent, type, and location of a child's abnormalities. Doctors classify CP according to the type of movement disorder involved — *spastic* (stiff muscles), *athetoid* (writhing movements), or *ataxic* (poor balance and coordination) — plus any additional symptoms, such weakness *(paresis)* or paralysis (*plegia)*. For example, *hemiparesis* (*hemi* = half) indicates that only one side of the body is weakened. *Quadriplegia* (*quad* = four) means all four limbs are afffected.

Spastic cerebral palsy is the most common type of the disorder. People have stiff muscles and awkward movements. Forms of spastic cerebral palsy include:

- *Spastic hemiplegia/ hemiparesis* typically affects the arm and hand on one side of the body, but it can also include the leg. Children with spastic hemiplegia generally walk later and on tip-toe because of tight heel tendons. The arm and leg of the affected side are frequently shorter and thinner. Some



The most common forms of cerebral palsy use Latin terms to describe the location or number of affected limbs, combined with the words plegia (paralysis) or paresis (weak).

children will develop an abnormal curvature of the spine (*scoliosis*). A child with spastic hemiplegia may also have seizures. Speech will be delayed and, at best, may be competent, but intelligence is usually normal.

- *Spastic diplegia/diparesis* involves muscle stiffness that is predominantly in the legs and less severely affects the arms and face, although the hands may be clumsy. Tendon reflexes in the legs are hyperactive. Toes point up when the bottom of the foot is stimulated. Tightness in certain leg muscles makes the legs move like the arms of a scissor. Children may require a walker or leg braces. Intelligence and language skills are usually normal.
- *Spastic quadriplegia/quadriparesis* is the most severe form of cerebral palsy and is often associated with moderate-to-severe intellectual disability. It is caused by widespread damage to the brain or significant brain malformations. Children will often have severe stiffness in their limbs but a floppy neck. They are rarely able to walk. Speaking and being understood are difficult. Seizures can be frequent and hard to control.

Dyskinetic cerebral palsy (also includes athetoid, choreoathetoid, and dystonic cerebral palsies) is characterized by slow and uncontrollable writhing or jerky movements of the hands, feet, arms, or legs. Hyperactivity in the muscles of the face and tongue makes some children grimace or drool. They find it difficult to sit straight or walk. Some children have problems hearing, controlling their breathing, and/or coordinating the muscle movements required

for speaking. Intelligence is rarely affected in these forms of cerebral palsy.

Ataxic cerebral palsy affects balance and depth perception. Children with ataxic CP will often have poor coordination and walk unsteadily with a wide-based gait. They have difficulty with quick or precise movements, such as writing or buttoning a shirt, or a hard time controlling voluntary movement such as reaching for a book.

Mixed types of cerebral palsy refer to symptoms that don't correspond to any single type of CP but are a mix of types. For example, a child with mixed CP may have some muscles that are too tight and others that are too relaxed, creating a mix of stiffness and floppiness.

What other conditions are associated with cerebral palsy?

Abnormal sensations and perceptions. Some individuals with CP experience pain or have difficulty feeling simple sensations, such as touch.

Contractures. Muscles can become painfully fixed into abnormal positions, called *contractures*, which can increase muscle spasticity and joint deformities in people with CP.

Delayed growth and development. Children with moderate to severe CP, especially those with spastic quadriparesis, often lag behind in growth and development. In babies this lag usually takes the form of too little weight gain. In young children it

can appear as abnormal shortness, and in teenagers it may appear as a combination of shortness and lack of sexual development. The muscles and limbs affected by CP tend to be smaller than normal, especially in children with spastic hemiplegia, whose limbs on the affected side of the body may not grow as quickly or as long as those on the normal side.

Dental problems. Many children with CP are at risk of developing gum disease and cavities because of poor dental hygiene. Certain medications, such as seizure drugs, can exacerbate these problems.

Drooling. Some individuals with CP drool because they have poor control of the muscles of the throat, mouth, and tongue.

Hearing loss. Impaired hearing is also more frequent among those with CP than in the general population. Some children have partial or complete hearing loss, particularly as the result of jaundice or lack of oxygen to the developing brain.

Impaired vision. Many children with CP have strabismus, commonly called "cross eyes," which left untreated can lead to poor vision in one eye and can interfere with the ability to judge distance. Some children with CP have difficulty understanding and organizing visual information. Other children may have defective vision or blindness that blurs the normal field of vision in one or both eyes.

Inactivity. Childhood inactivity is magnified in children with CP due to impairment of the motor centers of the brain that produce and control voluntary movement. While children with CP may exhibit increased energy expenditure during activities

of daily living, movement impairments make it difficult for them to participate in sports and other activities at a level of intensity sufficient to develop and maintain strength and fitness. Inactive adults with disability exhibit increased severity of disease and reduced overall health and well-being.

Incontinence. A possible complication of CP is incontinence, caused by poor control of the muscles that keep the bladder closed.

Infections and long-term illnesses. Many adults with CP have a higher risk of heart and lung disease, and pneumonia (often from inhaling bits of food into the lungs), than those without the disorder.

Intellectual disability. Approximately 30 – 50 percent of individuals with CP will be intellectually impaired. Mental impairment is more common among those with spastic quadriplegia than in those with other types of cerebral palsy.

Learning difficulties. Children with CP may have difficulty processing particular types of spatial and auditory information. Brain damage may affect the development of language and intellectual functioning.

Malnutrition. Swallowing, sucking, or feeding difficulties can make it difficult for many individuals with CP, particularly infants, to get proper nutrition and gain or maintain weight.

Seizure disorder. As many as half of all children with CP have one or more seizures. Children with both cerebral palsy and epilepsy are more likely to have intellectual disability.

Although symptoms may change over time, CP is not progressive.

Spinal deformities and osteoarthritis. Deformities of the spine — curvature (*scoliosis*), humpback (*kyphosis*), and saddle back *(lordosis*) — are associated with CP. Spinal deformities can make sitting, standing, and walking difficult and cause chronic back pain. Pressure on and misalignment of the joints may result in osteoporosis (a breakdown of cartilage in the joints and bone enlargement).

Speech and language disorders. Speech and language disorders, such as difficulty forming words and speaking clearly, are present in more than a third of persons with CP. Poor speech impairs communication and is often interpreted as a sign of cognitive impairment, which can be very frustrating to children with CP, especially the majority who have average to above average intelligence,

How is cerebral palsy diagnosed?

Most children with cerebral palsy are diagnosed during the first 2 years of life. But if a child's symptoms are mild, it can be difficult for a doctor to make a reliable diagnosis before the age of 4 or 5.

Doctors will order a series of tests to evaluate the child's motor skills. During regular visits, the doctor will monitor the child's development, growth, muscle tone, age-appropriate motor control, hearing and vision, posture, and coordination, in order to rule out other disorders that could cause similar symptoms. Although symptoms may change over time, CP is not progressive. If a child is

continuously losing motor skills, the problem more likely is a condition other than CP — such as a genetic or muscle disease, metabolism disorder, or tumors in the nervous system.

Lab tests can identify other conditions that may cause symptoms similar to those associated with CP.

Neuroimaging techniques that allow doctors to look into the brain (such as an MRI scan) can detect abnormalities that indicate a potentially treatable movement disorder. Neuroimaging methods include:

- **Cranial ultrasound** uses high-frequency sound waves to produce pictures of the brains of young babies. It is used for high-risk premature infants because it is the least intrusive of the imaging techniques, although it is not as successful as computed tomography or magnetic resonance imaging at capturing subtle changes in white matter — the type of brain tissue that is damaged in CP.
- **Computed tomography (CT)** uses x-rays to create images that show the structure of the brain and the areas of damage.
- **Magnetic resonance imaging (MRI)** uses a computer, a magnetic field, and radio waves to create an anatomical picture of the brain's tissues and structures. MRI can show the location and type of damage and offers finer levels of details than CT.

Another test, an **electroencephalogram**, uses a series of electrodes that are either taped or temporarily pasted to the scalp to detect electrical activity in the brain. Changes in the normal electrical pattern may help to identify epilepsy.

Some metabolic disorders can masquerade as CP. Most of the childhood metabolic disorders have characteristic brain abnormalities or malformations that will show up on an MRI.

Other types of disorders can also be mistaken for CP or can cause specific types of CP. For example, coagulation disorders (which prevent blood from clotting or lead to excessive clotting) can cause prenatal or perinatal strokes that damage the brain and produce symptoms characteristic of CP, most commonly hemiparetic CP. Referrals to specialists such as a child neurologist, developmental pediatrician, ophthalmologist, or otologist aid in a more accurate diagnosis and help doctors develop a specific treatment plan.

How is cerebral palsy treated?

Cerebral palsy can't be cured, but treatment will often improve a child's capabilities. Many children go on to enjoy near-normal adult lives if their disabilities are properly managed. In general, the earlier treatment begins, the better chance children have of overcoming developmental disabilities or learning new ways to accomplish the tasks that challenge them.

There is no standard therapy that works for every individual with cerebral palsy. Once the diagnosis is made, and the type of CP is determined, a team of health care professionals will work with a child and his or her parents to identify specific impairments and needs, and then develop an appropriate plan

to tackle the core disabilities that affect the child's quality of life.

Physical therapy, usually begun in the first few years of life or soon after the diagnosis is made, is a cornerstone of CP treatment. Specific sets of exercises (such as resistive, or strength training programs) and activities can maintain or improve muscle strength, balance, and motor skills, and prevent contractures. Special braces (called orthotic devices) may be used to improve mobility and stretch spastic muscles.

Occupational therapy focuses on optimizing upper body function, improving posture, and making the most of a child's mobility. Occupational therapists help individuals address new ways to meet everyday activities such as dressing, going to school, and participating in day-to-day activities.

The treatment team for children with cerebral palsy may include an occupational therapist to teach the skills necessary for day-to-day living. Above, an 18-month-old boy learns to feed himself.

Recreation therapy encourages participation in art and cultural programs, sports, and other events that help an individual expand physical and cognitive skills and abilities. Parents of children who participate in recreational therapies usually notice an improvement in their child's speech, self-esteem, and emotional well-being.

Speech and language therapy can improve a child's ability to speak, more clearly, help with swallowing disorders, and learn new ways to

communicate — using sign language and/or special communication devices such as a computer with a voice synthesizer, or a special board covered with symbols of everyday objects and activities to which a child can point to indicate his or her wishes.

Speech and language therapy can help treat the communication disorders associated with cerebral palsy.

Treatments for problems with eating and drooling are often necessary when children with CP have difficulty eating and drinking because they have little control over the muscles that move their mouth, jaw, and tongue. They are also at risk for breathing food or fluid into the lungs, as well as for malnutrition, recurrent lung infections, and progressive lung disease.

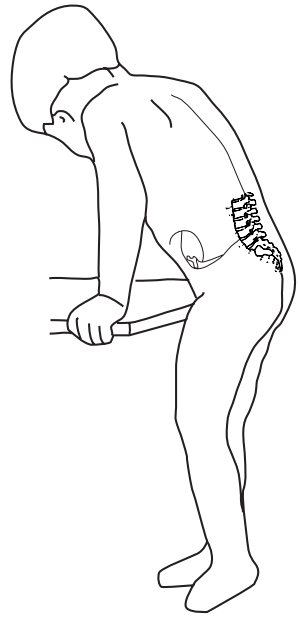
Drug treatments

Oral medications such as diazepam, baclofen, dantrolene sodium, and tizanidine are usually used as the first line of treatment to relax stiff, contracted, or overactive muscles. Some drugs have some risk side effects such as drowsiness, changes in blood pressure, and risk of liver damage that require continuous monitoring. Oral medications are most appropriate for children who need only mild reduction in muscle tone or who have widespread spasticity.

- Botulinum toxin (BT-A), injected locally, has become a standard treatment for overactive muscles in children with spastic movement disorders such as CP. BT-A relaxes contracted muscles by keeping nerve cells from over-activating muscle. The relaxing effect of a

BT-A injection lasts approximately 3 months. Undesirable side effects are mild and short-lived, consisting of pain upon injection and occasionally mild flu-like symptoms. BT-A injections are most effective when followed by a stretching program including physical therapy and splinting. BT-A injections work best for children who have some control over their motor movements and have a limited number of muscles to treat, none of which is fixed or rigid.

- Intrathecal baclofen therapy uses an implantable pump to deliver baclofen, a muscle relaxant, into the fluid surrounding the spinal cord. Baclofen decreases the excitability of nerve cells in the spinal cord, which then reduces muscle spasticity throughout the body. The pump can be adjusted if muscle tone is worse at certain times of the day or night. The baclofen pump is most appropriate for individuals with chronic, severe stiffness or uncontrolled muscle movement throughout the body.



The intrathecal baclofen pump system (shown above) delivers the muscle relaxant to the fluid surrounding the spinal cord. Baclofen decreases the excitability of nerve cells in the spinal cord, reducing muscle spasticity throughout the body.

Surgery

Orthopedic surgery is often recommended when spasticity and stiffness are severe enough to make walking and moving about difficult or painful. For many people with CP, improving the appearance of how they walk — their gait — is also important. Surgeons can lengthen muscles and tendons that are proportionately too short, which can improve mobility and lessen pain. Tendon surgery may help the symptoms for some children with CP but could also have negative long-term consequences. Orthopedic surgeries may be staggered at times appropriate to a child's age and level of motor development. Surgery can also correct or greatly improve spinal deformities in people with CP. Surgery may not be indicated for all gait abnormalities and the surgeon may request a quantitative *gait analysis* before surgery.

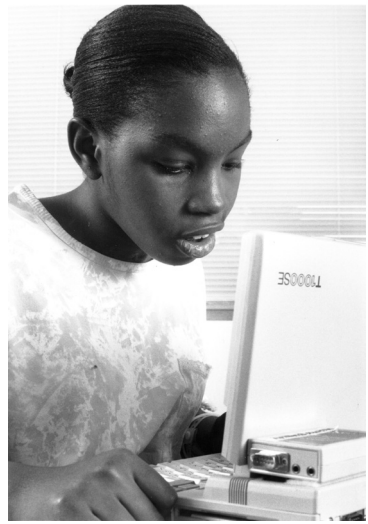
Surgery to cut nerves. *Selective dorsal rhizotomy* (SDR) is a surgical procedure recommended for cases of severe spasticity when all of the more conservative treatments — physical therapy, oral medications, and intrathecal baclofen — have failed to reduce spasticity or chronic pain. A surgeon locates and selectively severs overactivated nerves at the base of the spinal column. SDR is most commonly used to relax muscles and decrease chronic pain in one or both of the lower or upper limbs. It is also sometimes used to correct an overactive bladder. Potential side effects include sensory loss, numbness, or uncomfortable sensations in limb areas once supplied by the severed nerve.

Assistive devices

Assistive devices such devices as computers, computer software, voice synthesizers, and picture books can greatly help some individuals with CP improve communications skills. Other devices around the home or workplace make it easier for people with CP to adapt to activities of daily living.

Orthotic devices help to compensate for muscle imbalance and increase independent mobility. Braces and splints use external force to correct muscle abnormalities and improve function such as sitting or walking. Other orthotics help stretch muscles or the positioning of a joint. Braces, wedges, special chairs, and other devices can help people sit more comfortably and make it easier to perform daily functions.

Wheelchairs, rolling walkers, and powered scooters can help individuals who are not independently mobile. Vision aids include glasses, magnifiers, and large-print books and computer typeface. Some individuals with CP may need surgery to correct vision problems. Hearing aids and telephone amplifiers may help people hear more clearly.



Computers have made a big difference in the lives of people with cerebral palsy. Above, a teenager uses a computer to communicate and learn new words.

Complementary and alternative therapies

Many children and adolescents with CP use some form of complementary or alternative medicine. Controlled clinical trials involving some of the therapies have been inconclusive or showed no benefit and the therapies have not been accepted in mainstream clinical practice. Although there are anecdotal reports of some benefit in some children with CP, these therapies have not been approved by the U.S. Food and Drug Administration for the treatment of CP. Such therapies include hyperbaric oxygen therapy, special clothing worn during resistance exercise training, certain forms of electrical stimulation, assisting children in completing certain motions several times a day, and specialized learning strategies. Also, dietary supplements, including herbal products, may interact with other products or medications a child with CP may be taking or have unwanted side effects on their own. Families of children with CP should discuss all therapies with their doctor.

Stem cell therapy is being investigated as a treatment for cerebral palsy, but research is in early stages and large-scale clinical trials are needed to learn if stem cell therapy is safe and effective in humans. Stem cells are capable of becoming other cell types in the body. Scientists are hopeful that stem cells may be able to repair damaged nerves and brain tissues. Studies in the U.S. are examining the safety and tolerability of umbilical cord blood stem cell infusion in children with CP.

Are there treatments for other conditions associated with cerebral palsy?

Epilepsy. Many children with intellectual disability and CP also have epilepsy. In general, drugs are prescribed based on the type of seizures an individual experiences, since no one drug controls all types. Some individuals may need a combination of two or more drugs to achieve good seizure control.

Incontinence. Medical treatments for incontinence include special exercises, biofeedback, prescription drugs, surgery, or surgically implanted devices to replace or aid muscles.

Osteopenia. Children with CP who are unable to walk risk developing poor bone density (*osteopenia)*, which makes them more likely to break bones. In a study of older Americans funded by the National Institutes of Health (NIH), a family of drugs called *bisphosphonates*, which has been approved by the FDA to treat mineral loss in the elderly, also appeared to increase bone mineral density. Doctors may choose to selectively prescribe the drug off-label to children to prevent osteopenia.

Pain. Pain can be a problem for people with CP due to spastic muscles and the stress and strain on parts of the body that are compensating for muscle abnormalities. Some individuals may also have frequent and irregular muscle spasms that can't be predicted or medicated in advance. Diazepam can reduce the pain associated with muscle spasms and gabapentin has been used successfully to decrease the severity and frequency of painful spasms. Botulinum toxin injections have also been shown to decrease spasticity and pain. Intrathecal baclofen

has shown good results in reducing pain. Some children and adults have been able to decrease pain by using noninvasive and drug-free interventions such as distraction, relaxation training, biofeedback, and therapeutic massage.

Do adults with cerebral palsy face special health challenges?

Premature aging. The majority of individuals with CP will experience some form of premature aging by the time they reach their 40s because of the extra stress and strain the disease puts upon their bodies. The developmental delays that often accompany CP keep some organ systems from developing to their full capacity and level of performance. As a consequence, organ systems such as the cardiovascular system (the heart, veins, and arteries) and pulmonary system (lungs) have to work harder and they age prematurely.

Functional issues at work. The day-to-day challenges of the workplace are likely to increase as an employed individual with CP reaches middle age. Some individuals will be able to continue working with accommodations such as an adjusted work schedule, assistive equipment, or frequent rest periods.

The amount of emotional support someone has, how successful they are a coping with disappointment and stress, and whether or not they have an optimistic outlook about the future all have a significant impact on mental health.

Depression. Mental health issues can also be of concern as someone with cerebral palsy grows older. The rate of depression is three to four times higher in people with disabilities such as cerebral palsy. It appears to be related not so much to the severity of their disabilities, but to how well they cope with them. The amount of emotional support someone has, how successful they are at coping with disappointment and stress, and whether or not they have an optimistic outlook about the future all have a significant impact on mental health.

Post-impairment syndrome. This syndrome is marked by a combination of pain, fatigue, and weakness due to muscle abnormalities, bone deformities, *overuse syndromes* (sometimes also called repetitive motion injuries), and arthritis. Fatigue is often a challenge, since individuals with CP may use up to three to five times the amount of energy that able-bodied people use when they walk and move about.

Osteoarthritis and degenerative arthritis. Musculoskeletal abnormalities that may not produce discomfort during childhood can cause pain in adulthood. For example, the abnormal relationships between joint surfaces and excessive joint compression can lead to the early development of painful osteoarthritis and degenerative arthritis. Individuals with CP also may have limited strength and restricted patterns of movement, which puts them at risk for overuse syndromes and *nerve entrapments*.

Pain. Individuals with CP may have pain that can be acute (usually comes on quickly and lasts a short while) or chronic, and is experienced most commonly in the hips, knees, ankles, and the upper and lower

back. Individuals with spastic CP may have an increased number of painful sites and worse pain than those with other types of cerebral palsy. Preventive treatment aimed at correcting skeletal and muscle abnormalities early in life may help to avoid the progressive accumulation of stress and strain that causes pain. Dislocated hips, which are particularly likely to cause pain, can be surgically repaired.

Other medical conditions. Adults have higher than normal rates of other medical conditions secondary to their cerebral palsy, such as hypertension, incontinence, bladder dysfunction, and swallowing difficulties. Scoliosis is likely to progress after puberty, when bones have matured into their final shape and size. People with CP also have a higher incidence of bone fractures, occurring most frequently during physical therapy sessions.

What research is being done?

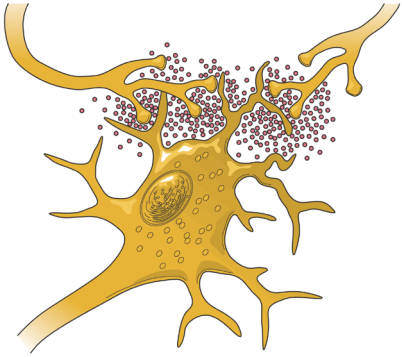
The National Institute of Neurological Disorders and Stroke, (NINDS), a part of the National Institutes of Health (NIH), is the nation's leading funder of basic, clinical, and translational research on brain and nervous system disorders. Another NIH agency, the *Eunice Kennedy Shriver* National Institute of Child Health and Human Development (NICHD), also conducts and supports research on cerebral palsy.

Much of what we now know about CP came from research sponsored by the NINDS, including the identification of new causes and risk factors for cerebral palsy, the discovery of drugs to control stiff

and spastic muscles and more precise methods to deliver them, refined surgical techniques to correct abnormalities in muscle and bone, and a greater understanding of how and why brain damage at critical stages of fetal development causes CP.

Many scientists think that a significant number of children develop CP because of mishaps early in **brain development**. They are examining how neurons (nerve cells) in the brain specialize and form the right connections with other brain cells, and they are looking for ways to prevent the factors that disrupt the normal processes of brain development.

Genetic defects are sometimes responsible for the brain malformations and abnormalities that cause cerebral palsy. Scientists are searching for the genes responsible for these abnormalities by collecting DNA samples from people with cerebral palsy and their families and using genetic screening techniques to discover linkages between individual genes and specific types of abnormality — primarily those associated with the process in the developing brain in which neurons migrate from where they are born to where they settle into neural circuits (called neural migration).



The brain chemical glutamate is necessary to help neurons communicate. But too much glutamate — called glutamate toxicity — overexcites and kills neurons. In the search for causes of cerebral palsy, scientists are studying conditions that result in high levels of glutamate in the brain.

Scientists are scrutinizing events in newborn babies' brains, such as bleeding, epileptic seizures, and

breathing and circulation problems, which can cause the **abnormal release of chemicals** that triggers the kind of damage that causes cerebral palsy. For example, research has shown that bleeding in the brain unleashes dangerously high amounts of glutamate, a chemical that helps neurons communicate. However, too much glutamate overexcites and kills neurons. By learning how brain chemicals that are normally helpful become dangerously toxic, scientists will have opportunities to develop new drugs to block their harmful effects.

Researchers are using **imaging techniques** and neurobehavioral tests to predict those preterm infants who will develop cerebral palsy. If these screening techniques are successful, doctors will be able to identify infants at risk for cerebral palsy before they are born.

Periventricular **white matter damage** — the most common cause of CP — is characterized by death of the white matter around the fluid-filled ventricles in the brain. The periventricular area contains nerve fibers that carry messages from the brain to the body's muscles. NINDS-sponsored researchers are hoping to develop preventative strategies for white matter damage. For example, researchers are examining the role the brain chemicals play on white matter development in the brain. Another NINDS-funded project involves the development of a novel mouse model and cell-based therapies for perinatal white matter injury. Researchers funded by NINDS are studying a chemical found naturally in the body, called erythropoietin to see if it decreases the risk of CP in prematurely born infants.

One type of physical therapy called constraint-resistant therapy — which involves restraining the stronger arm and forcing the weaker arm to perform intensive activities — has been shown effective for some children with cerebral palsy.

NIH-funded scientists continue to look at new therapies and novel ways to use existing options to treat individuals with CP, including:

Constraint-induced therapy (CIT) is a promising therapy for CP. CIT typically involves restraining the stronger limb (such as the "good" arm in a person who has been affected by a stroke on one side of the body) in a cast and forcing the weaker arm to perform intensive activities every day over a period of weeks. A clinical study sponsored by the NICHD is examining the use of different dosage levels of daily training using either full-time cast immobilization vs. part-time splint restraint in improving upper body extremity skills in children with weakness on both sides of their body. Study findings will establish evidence-based practice standards to improve lifelong neuromotor capacity in individuals with CP.

Functional electrical stimulation (FES) — the therapeutic use of low-level electrical current to stimulate muscle movement and restore useful movements such as standing or stepping — is an effective way to target and strengthen spastic muscles. Researchers are evaluating how FES-assisted stationary cycling can improve physical conditioning and general lower extremity muscle strength in adolescents. ***Robotic therapy*** that

applies controlled force to the leg during the swing phase of gait is may improve the efficacy of body weight supported treadmill training in children with CP. The results from this NICHD study will lead to an innovative clinical therapy aimed at improving locomotor function in children with CP.

Botulinum toxin (Botox), injected locally, has become a standard treatment in children with spastic movement disorders such as CP. Recent animal studies suggest Botox degrades bone but there are no studies of its skeletal consequences in humans. Other research shows a low intensity vibration treatment can improve bone structure in the lower extremity leg bones of children with CP. In a novel clinical study being conducted by NICHD, researchers are determining the effect of Botox treatment in conjunction with a daily vibration treatment on bone mass and bone structure in children with spastic CP.

Systemic hypothermia — the controlled medical cooling of the body's core temperature — appears to protect the brain and decrease the rate of death and disability from certain disorders and brain injuries. Previous studies have shown that hypothermia is effective in treating neurologic symptoms in term or late preterm babies less than one month old that are attributed to hypoxic-ischemia (HIE, brain injury due to a severe decrease in the oxygen supply to the body), which can cause quadriplegic CP, with or without movement disorder. In an effort to determine the most effective cooling strategies, NICHD-funded researchers are studying different cooling

treatments to improve the chance of survival and neurodevelopment outcomes 18-22 months post-treatment in infants with neurologic symptoms attributed to HIE. Other researchers are examining if combined therapy using hypothermia and recombinant erythropoietin (a hormone that promotes the growth of new red blood cells and increases oxygen levels in the blood) is more effective than either therapy alone in treating neurodevelopmental handicaps in an animal model involving lack of oxygen before, during, or just after birth.

As researchers continue to explore new treatments for cerebral palsy and to expand our knowledge of brain development, we can expect significant improvements in the care of children with cerebral palsy and many other disorders that strike in early life.

Glossary

acquired cerebral palsy — cerebral palsy that occurs as a result of injury to the brain after birth or during early childhood.

Apgar score — a numbered scoring system doctors use to assess a baby's physical state at the time of birth.

asphyxia — a lack of oxygen due to trouble with breathing or poor oxygen supply in the air.

ataxia — the loss of muscle control.

athetoid — making slow, sinuous, involuntary, writhing movements, especially with the hands.

bilirubin — a bile pigment produced by the liver of the human body as a byproduct of digestion.

bisphosphonates — a family of drugs that strengthen bones and reduce the risk of bone fracture in elderly adults.

botulinum toxin — a drug commonly used to relax spastic muscles; it blocks the release of acetylcholine, a neurotransmitter that energizes muscle tissue.

cerebral — relating to the two hemispheres of the human brain.

cerebral dysgenesis — defective brain development.

choreoathetoid — a condition characterized by aimless muscle movements and involuntary motions.

congenital cerebral palsy — cerebral palsy that is present at birth from causes that have occurred during fetal development.

contracture — a condition in which muscles become fixed in a rigid, abnormal position, which causes distortion or deformity.

developmental delay — behind schedule in reaching the milestones of early childhood development.

dyskinetic — the impairment of the ability to perform voluntary movements, which results in awkward or incomplete movements.

dystonia (dystonic) — a condition of abnormal muscle tone.

gait analysis — a technique that uses cameras, force plates, electromyography, and computer analysis to objectively measure an individual's pattern of walking.

gestation — the period of fetal development from the time of conception until birth.

hemiparesis — paralysis affecting only one side of the body.

hypertonia — increased muscle tone.

hypotonia — decreased muscle tone.

hypoxic-ischemic encephalopathy — brain damage caused by poor blood flow or insufficient oxygen supply to the brain.

intracranial hemorrhage — bleeding in the brain.

***intrathecal baclofen* —** baclofen that is injected into the cerebrospinal fluid of the spinal cord to reduce spasticity.

***jaundice* —** a blood disorder caused by the abnormal buildup of bilirubin in the bloodstream.

***kyphosis* —** a humpback-like outward curvature of the upper spine.

***lordosis* —** an increased inward curvature of the lower spine.

***orthotic devices* —** special devices, such as splints or braces, used to treat posture problems involving the muscles, ligaments, or bones.

***osteopenia* —** reduced density and mass of the bones.

***palsy* —** paralysis, or the lack of control over voluntary movement.

***-paresis or -plegia* —** weakness or paralysis. In cerebral palsy, these terms are typically combined with other phrases that describe the distribution of paralysis and weakness; for example, quadriplegia means paralysis of all four limbs.

***periventricular leukomalacia (PVL)* —** "peri" means near; "ventricular" refers to the ventricles or fluid spaces of the brain; and "leukomalacia" refers to softening of the white matter of the brain. PVL is a condition in which the cells that make up white matter die near the ventricles. Under a microscope, the tissue looks soft and sponge-like.

***placenta* —** an organ that joins a mother with her unborn baby and provides nourishment and sustenance.

***quadriplegia* —** paralysis of both the arms and legs.

***Rh incompatibility* —** a blood condition in which antibodies in a pregnant woman's blood attack fetal blood cells and impair an unborn baby's supply of oxygen and nutrients.

***rubella* —** (also known as German measles) a viral infection that can damage the nervous system of an unborn baby if a mother contracts the disease during pregnancy.

***scoliosis* —** a disease of the spine in which the spinal column tilts or curves to one side of the body.

***selective dorsal rhizotomy* —** a surgical procedure in which selected nerves are severed to reduce spasticity in the legs.

***spastic (or spasticity)* —** describes stiff muscles and awkward movements.

***spastic diplegia (or diparesis)* —** a form of cerebral palsy in which spasticity affects both legs, but the arms are relatively or completely spared.

***spastic hemiplegia (or hemiparesis)* —** a form of cerebral palsy in which spasticity affects an arm and leg on one side of the body.

***spastic quadriplegia (or quadriparesis)* —** a form of cerebral palsy in which all four limbs are paralyzed or weakened equally.

***tremor* —** an involuntary trembling or quivering.

Credits

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National Institute of
Neurological Disorders
and Stroke

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NIH Publication No. 13-159
August 2013