

CEREBRAL PALSY

WHAT IS THIS CONDITION

We do not know the cause of most cases of cerebral palsy. That is, we are unable to determine what caused cerebral palsy in most children who have congenital CP. We do know that the child who is at highest risk for developing CP is the premature, very small baby who does not cry in the first five minutes after delivery, who needs to be on a ventilator for over four weeks, and who has bleeding in his brain. Babies who have congenital malformations in systems such as the heart, kidneys, or spine are also more likely to develop CP, probably because they also have malformations in the brain. Seizures in a newborn also increase the risk of CP. There is no combination of factors which always results in an abnormally functioning individual. That is, even the small premature infant has a better than 90 percent chance of not having cerebral palsy. There are a surprising number of babies who have very stormy courses in the newborn period and go on to do very well. In contrast, some infants who have rather benign beginnings are eventually found to have severe mental retardation or learning disabilities.

CEREBRAL PALSY IN THE NEWBORN

Many children with cerebral palsy have a congenital malformation of the brain, meaning that the malformation existed at birth and was not caused by factors occurring during the birthing process. Not all of these malformations can be seen by the physician, even with today's most sophisticated scans, but when CP is recognized in a newborn, a congenital malformation is suspected. When a diagnosis of CP is made, the mother and father often feel guilty and wonder what they did to cause their child to have this disorder. While it is certainly true that good prenatal care is an essential part of preventing congenital problems, it must be stated that congenital problems, or "birth defects," often occur even when the mother has strictly followed her physician's advice in caring for herself and the developing infant. Though the

causes of "birth defects" are usually unknown, we do know that the developing brain can be affected by several factors. When the fetus is exposed to certain chemicals or infections through the expectant mother, for example. The developing brain can be injured if the expectant

mother suffers severe physical trauma, the fetal brain can be injured, too, but this is rare. Finally, prematurity and a low birth weight have been shown to be related to an increased incidence of specific disorders. Many chemicals are known to adversely affect the developing brain, alcohol being the most commonly used. The term Fetal Alcohol Syndrome describes the long-term, multi-system effect of alcohol on a child whose mother abused alcohol during the pregnancy. When a fetus is exposed to large amounts of alcohol, several body systems, including the neurological system will almost certainly suffer damage. Cigarette smoking by the mother has been shown to decrease birth weight, and low birth weight is associated with several disorders, including cerebral palsy. Severe malnutrition in the mother can adversely affect brain growth in the fetus, and it, too, can result in a low birth weight. The use of cocaine or crack by the expectant mother is associated with blood vessel complications, and these complications affect many organs as well as the central nervous system. Cocaine use is increasing and thus becoming more prevalent as cause of brain damage in infants. Most infants whose mothers used cocaine during pregnancy develop mental retardation rather than cerebral palsy, however. Infections such as rubella (German measles), toxoplasmosis, and cytomegalovirus (CMV), (if a woman has them during pregnancy), also may injure the brain of the fetus. Rubella can be prevented by immunization, prior to becoming pregnant, and the chances of becoming infected with toxoplasmosis can be minimized by not handling the feces of cats and by avoiding raw or uncooked meat.

Congenital infection with human immunodeficiency virus (HIV, the virus that causes AIDS) also causes brain damage in children, though it usually causes mental retardation rather than CP. It is likely that many other infections in the expectant mother injure the developing fetus, but they are not recognized as causative factors because the woman who has the infection either does not recognize the symptoms of infection or is symptom-free. Premature infants are at a much higher risk for developing cerebral palsy than full-term babies, and the risk increases

as the birth weight decreases. Between 5 and 8 percent of infants weighing less than 1500 grams (3 pounds) at birth develop cerebral palsy, and infants weighing less than 1500 grams are 25 times more likely to develop cerebral palsy than infants who are born at full term weighing more than 2500 grams.

any premature infants suffer bleeding within the brain, called intraventricular hemorrhages, intracranial hemorrhages. Again, the highest frequency of hemorrhages is found in the babies with the lowest weight: the problem is rare in babies who weigh more than 2000 grams (4 pounds). This bleeding may damage the part of the brain that controls motor function and thereby lead to cerebral palsy. If the hemorrhage results in destruction of normal brain tissue (a condition called periventricular leukomalacia) and small cysts around the ventricles and in the motor region of the brain, then that infant is more likely to have CP than an infant with hemorrhages alone. Does prematurity "cause" cerebral palsy, or do some infants who are born prematurely have abnormal brains from the beginning, leading to their premature births? We do not know the answer to this question.

CEREBRAL PALSY FROM THE BIRTHING PROCESS

There are no specific events that, if they occur during pregnancy, delivery, or infancy, will always occurring at birth or right after birth). This is apparently why the incidence of CP in undeveloped and poverty stricken areas of the world, where infant mortality is very high, is the same as in northern Europe, where infant mortality is the lowest. It also explains why modern obstetrical care, including monitoring and a high rate of Cesarean section, has lowered infant mortality rates but not the incidence of cerebral palsy. One large study, for example, has shown that more than 60 percent of all pregnancies have at least one complication, and that most of these complications cause no problems. For instance, 25 percent of all newborns have the umbilical cord wrapped around their neck, and 16 percent passed meconium (had the first bowel movement) at the time of birth. These "birth events" and the development of CP have only a small correlation. In other words, the chances of a child developing CP were nearly the same whether the child was born with a cord wrapped around her neck or not. On the

other hand, newborns in this study who had very low Apgar scores (less than 3 at 20 minutes) had a risk 250 times greater than infants with normal Apgar scores of developing cerebral palsy. An Apgar score at this level suggests that the infant suffered severe asphyxia (lack of sufficient oxygen to the brain) during birth. Half of the infants who suffered severe asphyxia during birth did not develop cerebral palsy, however. When CP is diagnosed in childhood, it is often discovered that the child suffered asphyxia at birth, but the asphyxia is usually considered the symptom of an otherwise sick baby with a neurological problem, and not the primary cause of CP. In two different large studies, only about 9 percent of children with CP were thought to have CP directly and exclusively related to asphyxia at delivery. Ninety-one percent of the babies had other inherent causes which led to prematurity or perinatal or neonatal problems (problems In the nineteenth century, Dr. William John Little described cerebral palsy and stated that the condition was due to birth injury in most cases. Cerebral palsy is also known as Little's disease and static encephalopathy, but the term cerebral palsy is most widely used. Dr. Sigmund Freud (who was a prominent neurologist before he founded the field of psychiatry) also investigated the causes of cerebral palsy. Freud thought that the condition was due to something which occurred before the child's birth. He argued that the problems seen at birth were often due to an abnormality present in the baby before birth, rather than being caused by the birthing process. This view of Freud's was greatly ignored in the first half of this century, but recent research has lent support to the idea that cerebral palsy is more often a result of a congenital abnormality than to an injury sustained at birth. Nevertheless, the birthing process can be traumatic for the infant, and injuries occurring during birth do sometimes cause cerebral palsy. Modern prenatal care and improved obstetric care have significantly reduced the incidence of birth injury, but it is unlikely that it will ever be completely eliminated.

CEREBRAL PALSY IN THE INFANT AND CHILD

During infancy and early childhood, the child is completely dependent on others for his or her safety and protection. Protecting the child from injury is one of the most important responsibilities of the

child's caregivers. One such injury is asphyxia, which can damage the brain in a variety of ways,

and is the number one cause of CP in this age group. The three most common causes of asphyxia in the young child are: choking on foreign objects such as toys and pieces of food (including peanuts, popcorn, and hot dogs); poisoning; and near drowning. The brain may also be damaged when it is physically traumatized as a result of a blow to the head. A child who falls or is involved in a motor vehicle accident or is the victim of physical abuse may suffer irreparable injury to the brain. One form of child abuse is the shaken baby syndrome, in which the caretaker is trying to quiet the baby by shaking too vigorously, causing the brain to strike repeatedly against the skull under high pressure.

Severe infections, especially meningitis or encephalitis, can also lead to brain damage in this age group. Meningitis is inflammation of the meninges (the covering of the brain and the spinal cord), usually caused by a bacterial infection, and encephalitis is brain inflammation which may be caused by bacterial or viral infections. Either of these infections can cause disabilities ranging from hearing loss to CP to severe retardation.

WHAT ARE SOME DISORDERS WHICH ARE NOT CEREBRAL PALSY BUT RESEMBLE CEREBRAL PALSY?

Children with disabilities have many problems in common, especially problems involving interactions with family members and society at large. The physical and medical problems of children with disabilities vary widely, however. Some of the problems caused by various disorders resemble those affecting children with cerebral palsy, but on closer inspection the medical issues turn out to be quite distinct.

Children with spinal cord dysfunction, for example, face medical problems such as insensate skin and bowel and bladder dysfunction, which differ markedly from the medical problems faced by children with cerebral palsy. Spinal cord dysfunction may be a result of spinal cord injury, spina bifida (meningomyelocele), or a congenital spinal cord malformation. Another large group of children who at time may look similar to those with cerebral palsy are children with temporary

motor problems resulting from closed head injuries, seizures, drug overdoses, or some brain tumors. The medical issues for this group of children are also different

from the medical issues for children with cerebral palsy, because these injuries can occur at any age

and the severity of the problems caused by these injuries changes over time. We can also say that disorders that are primarily of muscle, nerve, and bone are not cerebral palsy by definition. Such conditions include muscular dystrophy, peripheral neuropathies such as Charcot-Marie-Tooth disease, and osteogenesis imperfecta. All of these conditions are associated with specific medical problems. Children with progressive neurologic disorders (including Rett's syndrome, leukodystrophy, and Tay-Sach's disease) also have medical needs which are different from those of children with cerebral palsy.

Some children with chromosomal anomalies (for example, trisomy 13 and 18) or congenital disorders (hereditary spastic paraplegia, for example) may appear similar to children with cerebral palsy; others, such as children with Down's syndrome, appear very different from children with cerebral palsy. Children with these disorders have some problems in common with children who have cerebral palsy; they also have problems that are unique for children with that specific disorder.

HOW IS A DIAGNOSIS OF CEREBRAL PALSY MADE?

Many of the normal developmental milestones, such as reaching for toys (3-4 months), sitting (6-7 months), and walking (10-14 months), are based on motor function. A physician may suspect cerebral palsy in a child whose development of these skills is delayed. In making a diagnosis of cerebral palsy, the physician takes into account the delay in developmental milestones as well as physical findings that might include abnormal muscle tone, abnormal movements, abnormal reflexes and persistent infantile reflexes. Making a definite diagnosis of cerebral palsy is not always easy, especially before the child's first birthday. In fact, diagnosing cerebral palsy usually involves a period of waiting for the definite and permanent appearance of specific motor problems. Most children with cerebral palsy can be diagnosed by the age of 18

months, but eighteen months is a long time for parents to wait for a diagnosis, and this is understandably a difficult period for them. Making a diagnosis of cerebral palsy is also difficult when, for example, a two-year-old has suffered a head injury. The child may immediately appear to be severely injured, and three months after the injury he may have symptoms that are typical of a child with cerebral palsy. But one year after the injury such a child may be completely normal. This child does not have cerebral palsy. Although he has a scar on his brain, the scar is not permanently impairing his motor activities. After injury, waiting and observing are necessary before the diagnosis can be made.

DO X-RAYS OR OTHER TESTS HELP IN DIAGNOSIS CEREBRAL PALSY?

As noted above, in making a diagnosis of cerebral palsy the most meaningful aspect of the examination is the physical evidence of abnormal motor function. A diagnosis of cerebral palsy cannot be made on the basis of an x-ray or blood test, though the physician may order such tests to exclude other neurologic diseases (such as those mentioned above). Blood tests and chromosome analysis are helpful in diagnosing hereditary conditions that may influence the parents' future child-bearing decisions. When the tests indicate that a child's condition is something other than cerebral palsy and that the condition is inherited, family members will benefit from genetic counselling. Cerebral palsy is not a hereditary condition, however, and these tests will neither establish nor rule out a diagnosis of CP.

Magnetic resonance imaging (MRI) and Computed Tomography (CT) scans are often ordered when the physician suspects that the child has cerebral palsy. These tests may provide evidence of hydrocephalus (an abnormal accumulation of fluid in the cerebral ventricles), and they may be used to exclude other causes of motor problems. These scans do not prove whether a child has a cerebral palsy; nor do they predict how a specific child will function as she grows. Thus, children with normal scans may have severe cerebral palsy, and children with clearly abnormal scans occasionally appear totally normal or have only mild physical evidence of cerebral palsy. As a group, though, children with cerebral palsy do have brain scars, cysts, and other changes which show

up on scans more frequently than in normal children. Therefore, when a scar is seen on a CT scan of the brain of a child whose physical examination suggests he may have cerebral palsy, the scar is one more piece of evidence indicating that the child is likely to have motor problems in the future.

WHAT ARE THE DIFFERENT TYPES OF CEREBRAL PALSY?

Cerebral palsy may be classified by the type of movement problem (such as spastic or athetoid cerebral palsy) or by the body parts involved (hemiplegia, diplegia, and quadriplegia). Spasticity refers to the inability of a muscle to relax, while athetosis refers to an inability to control the movement of a muscle. Infants who at first are hypotonic wherein they are very floppy may later develop spasticity. Hemiplegia is cerebral palsy that involves one arm and one leg on the same side of the body, whereas with diplegia the primary involvement is both legs. Quadriplegia refers to a pattern involving all four extremities as well as trunk and neck muscles. Another frequently used classification is ataxia, which refers to balance and coordination problems. The motor disability of a child with CP varies greatly from one child to another; thus generalizations about children with cerebral palsy can only have meaning within the context of the subgroups described above. For this reason, subgroups will be used in this book whenever treatment and outcome expectations are discussed. Most professionals who care for children with cerebral palsy understand these diagnoses and use them to communicate about a child's condition.

As noted above, a useful method for making subdivisions is determined by which parts of the body are involved. Although almost all children with cerebral palsy can be classified as having hemiplegia, diplegia, or quadriplegia, there are significant overlaps which have led to the use of additional terms, some of which are very confusing. To avoid confusion, most of the discussion in his book will be limited to the use of these three terms. Occasionally such terms as paraplegia, double hemiplegia, triplegia, and pentaplegia may occasionally be encountered by the reader; these classifications are also based on the parts of the body involved. The dominant type of movement or muscle coordination problem is the other method by which children are subdivided and

classified to assist in communicating about the problems of cerebral palsy. The component which seems to be causing the most problem is often used as the categorizing term. For example, the child with spastic diplegia has mostly spastic muscle problems, and most of the involvement is in the legs, but the child may also have a smaller component of athetosis and balance problems. The child with athetoid quadriplegia, on the other hand, would have involvement of both arms and legs, primarily with athetoid muscle problems, but such a child often has some ataxia and spasticity as well. Generally a child with quadriplegia is a child who is not walking independently. The reader may be familiar with other terms used to define specific problems of movement or muscle function terms such as: dystonia, tremor, ballismus, and rigidity. The words severe, moderate, and mild are also often used in combination with both anatomic and motor function classification terms (severe spastic diplegia, for example), but these qualifying words do not have any specific meaning. They are subjective words and their meaning varies depending on the person who is using them.

WHAT ARE THE RIGHT WORDS TO USE WHEN REFERRING TO CHILDREN WITH CEREBRAL PALSY?

Cerebral palsy is the term used to describe the motor impairment resulting from brain damage in the young child regardless of the cause of the damage or its effect on the child. Impairment is the correct term to use to define a deviation from normal, such as not being able to make a muscle move or not being able to control an unwanted movement. Disability is the term used to define a restriction in the ability to perform a normal activity of daily living which someone of the same age is able to perform. For example, a three year old child who is not able to walk has a disability because normal three year old can walk independently. Handicap is the term used to describe a child or adult who, because of the disability, is unable to achieve the normal role in society commensurate with his age and socio-cultural milieu. As an example, a sixteen-year- old who is unable to prepare his own meal or care for his own toileting or hygiene needs is handicapped. On the other hand, a sixteen-year- old who can walk only with the assistance of crutches but who attends a regular school and is fully independent in

activities of daily living is disabled but not handicapped. All disabled people are impaired, and all handicapped people are disabled, but a person can be impaired and not necessarily be disabled, and a person can be disabled without being handicapped. In the past there has been a disturbing lack of awareness and sensitivity, both among the general public and in literature, with respect to the words used when people with disabilities are discussed. But an increasing amount of attention is being paid to such language in our society along with issues of education, employment, and public access for disabled individuals. Certainly, the use of obviously pejorative expressions has always been inappropriate, and the formerly accepted practice of referring individuals by their disability ("the epileptic," "the spastic," "the retarded child") is no longer acceptable. While it may take years for our language to catch up with our changing views, the current acceptable terminology stresses the individual person and then mentions the disability that person has, therefore, we refer to a girl with spastic diplegia or a boy with mental retardation. Clearly, this language acknowledges that there is much more to this individual than his or her disability. Other terms that have recently come into use represent an even more enlightened view. For example, the child who is mentally challenged, rather than the child who is mentally retarded. In this book, we have chosen to use language that reflects the appropriate current societal goal of employing respectful terminology and that also reflects our concern with presenting information in a way that can be understood by the general reader. We fully realize that there may be newer, even

better terms to use, but we will not use them when there is a significant risk of introducing confusion into our discussion.

WHAT IS THE PROGNOSIS FOR THE CHILD WITH CEREBRAL PALSY?

The first questions usually asked by parents after they are told their child has cerebral palsy are "What will my child be like?" and "Will he walk?" Predicting what a young child with cerebral palsy will be like or what he will or will not do (called the prognosis) is very difficult. Any predictions for an infant under six months of age are little better than guesses, and even for children younger than one year it is often very

difficult to predict the pattern of involvement. By the time the child is two years old, however, the physician can determine whether the child has hemiplegia, diplegia, or quadriplegia. Based on this involvement pattern, some predictions can be made. It is worth saying again that children with cerebral palsy do not stop doing activities once they have begun to do them. Such a loss of skills, called regression, is not characteristic of cerebral palsy. If regression occurs, it is necessary to look for a different cause of the child's problems. In order for a child to be able to walk, some major events in motor control have to occur. A child must be able to hold up his head before he can sit up on his own, and he must be able to sit independently before he can walk on his own. It is generally assumed that if a child is not sitting up by himself by age 4 or walking by age 8, he will never be an independent walker. But a child who starts to walk at age 3 will certainly continue to walk and will be walking when he is 13 years old unless he has a disorder other than CP.

It is even more difficult to make early predictions of speaking ability or mental ability than it is to predict motor function. Here, too, evaluation is much more reliable after age 2, although a motor disability can make the evaluation of intellectual function quite difficult. Sometimes "motor-free" tests which can assess intellectual ability without, the person being tested, needing to use his hands are administered by psychologists who have expertise in their use. Overall, the intellectual ability of the person, far more than their physical disability, will determine the person's prognosis. In other words, mental retardation is far more likely than cerebral palsy to impair a child's ability to function.

WHAT CAN THE PHYSICIAN TELL THE PARENTS EARLY ON?

Parents are naturally concerned when their newborn child has problems, and physicians need to

evaluate the child's condition and prognosis as well as they can. For example, evidence of a bleed in the child's brain should be discussed with parents, although the outcome of such a bleed cannot be predicted. As we've discussed, the diagnosis of cerebral palsy cannot be made at

birth and, most assuredly, the extent and severity of involvement that an individual child might eventually have is

impossible to assess at birth. Many neonatologists, aware of the interaction that generally occurs between the newborn and parents, avoid discussing the child's problems in detail because they want to permit this interaction to take place. The presumption of a bleak future for a child sometimes causes parents to withdraw from the child and this can have a significant negative effect on the child. Physicians usually communicate their concerns in terms of the child's symptoms, such as muscle problems, and prepare parents for the possibility of neurologic damage. Clearly, it is part of the physician's role to inform parents, but the variability of outcome makes it virtually impossible for the physician to predict the future, and so the physician must weigh the need to inform (and the imprecision of information) against the need for the parents to have hope for, and to become close to their child.

HOW AGGRESSIVE SHOULD TREATMENT BE GIVEN A SICK NEWBORN?

Many times when a child is a few years old and severely disabled, parents begin to wonder whether treatment should have been less aggressive than it was. Given the tremendous uncertainties in outcome, physicians and parents usually choose to treat newborns and preserve life with the hope that the outcome will be a good one. There are clearly exceptions, such as when the baby has a known chromosomal defect (such as trisomy 18), where the poor prognosis is known and where very aggressive treatment may not be used. However, in the majority of cases the information regarding ultimate outcome is not available, and families and physicians do the best they can with the limited information they have. Often the prognosis is based on information from studies of a large number of babies with a similar birth weight. The chance of an individual baby having cerebral palsy or mental retardation (expressed as a percentage) is derived from these studies. Nevertheless, it is impossible to know whether an individual infant will fit into the 70-90 percent group that has a good outcome or the 10-30 percent group with a poor outcome.

The role of the physician is to gather as much information about the child's condition as possible and to convey this information to families along with the best information available about chances for outcome. The role of the family is to help in the decision-making process when there are decisions to be made about further aggressive treatment, though ultimately it is the physician's responsibility to decide what should or should not be done. Most physicians will take into consideration a family's wishes, but physicians cannot give up their legal and moral obligation to do what is best for their patient, nor can a physician withhold treatment without the family's permission. The problem is trying to figure out what is best. At the time the decisions must be made it is often very difficult to know what will ultimately be best. A decision to treat aggressively usually involves the use of sophisticated equipment, although availability of such technology does not mean that it must always be used, and there are clearly times when it is more humane to withhold or withdraw aggressive treatment. These are never easy decisions to make. Clergy, social workers, ethicists, and other health care workers who have come to know the patient and family often help in making a decision about what is best.

HOW CAN REALISTIC GOALS BE DESIGNED FOR THE CHILD WITH CEREBRAL PALSY?

When it comes to expectations and questions of what the future holds for the child with CP, it is important to maintain a combination of optimism and realism, just as one would with any child. Suppose, for example, that the parent of a non-disabled three year old has hopes and expectations that the child will go to college and law school, enter politics, and eventually become President of the United States. Some of these expectations are realistic and are likely to be met, while others are extremely unlikely to occur to the point of being clearly unrealistic. Regardless of these realistic and not realistic expectations, however, the parent needs to care for the child as a three year old and

not as a college student or as a politician. It is equally important for the parent of a child with

cerebral palsy to understand the child's present and future abilities. That parent's expectations are

also probably a combination of realistic and unrealistic goals for the child, but in time, with

professional help, the parent will develop a set of mostly realistic goals and it is to these goals that

the parent, child, and professional will dedicate their effort.

Occasionally, difficulties in communication arise when the parents, educators, and medical care providers discuss present abilities. As stated in the Preface, a significant goal of this book is to., improve this communication so that parents, educators, and medical care providers can communicate their impressions to each other regarding a specific child and in this way help the child function at his or her maximum ability. An attempt to define future expectations is usually most important in the teenage years and beyond, when function is better defined and the future looks more clear to everyone involved.

WHAT MEDICAL PROBLEMS ARE ASSOCIATED WITH CEREBRAL PALSY?

Children with cerebral palsy have many problems, not all of them related to the brain injury. Most

of these complications are nevertheless neurological. They include epilepsy, mental retardation,

learning disabilities, and attention deficit-hyperactivity disorder. These problems are discussed in

chapter 3, as are problems that occur less commonly, such as swallowing problems in children with

spastic quadriplegia. In the next chapter, we look at normal pediatric development, to establish a basis with which to compare the development and behavior of a child with cerebral palsy.

Children with cerebral palsy may also develop hip subluxation or have problems with the gait.