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Cerebral Palsy

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Cerebral palsy (CP) is a handicapping condition that can present with such vastly different expressions that it is easy to question the appropriateness of the term. Despite these misgivings, CP does exist. There are common understandings of its definition, a generally accepted classification system, increasing knowledge about its etiology, a body of knowledge about its epidemiology, and increasing numbers of treatment options.

CP is often described as a group of nonprogressive, but often changing, motor impairment syndromes secondary to lesions or anomalies of the brain arising at any time during early brain development (Mutch, Alberman, Hagberg, Kodama, & Perat, 1992). The tone abnormalities of CP can range from spasticity to hypotonicity, can be mixed, and can vary in one child throughout the day. An important component of the definition of CP is that it is nonprogressive. In fact, many children improve functionally over time consistent with the nature of pediatric neurologic maturation. Central to the definition of CP is the concept that it is a disorder of the brain and not of the musculoskeletal system. Understanding that CP is a neurodevelopmental disability with its primary impact on the motor system will assist families in understanding the impact of CP on a child's or adult's functional skills.

CP was first described in the medical literature in the late 19th century by Dr William Little. Little's description of the condition in his article, "On the Influence of Abnormal Parturition, Difficult Labours, Premature Birth . . . " set the stage for our understanding of CP as a condition that results from anoxia during labor and delivery (Little, 1862, article reprinted in (not alive in 1958) 1958). Unfortunately, a half century of epidemiologic research refuting that conclusion has not changed the lay perception that CP is caused by a bad labor and delivery experience with anoxic damage

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to the brain. While hypoxic ischemic encephalopathy occurs, it is only an infrequent cause of CP (Nelson & Grether, 1999).

Currently, as in the case for most developmental disabilities, a cure for CP is not available. The neurologic damage cannot be undone nor can a surgeon create new neuronal tissue. Nevertheless, medical professionals do provide assistance to the child and family through diagnosis, case management, and interventions that minimize impairment and disability and prevent secondary disabilities, as do related healthcare and rehabilitation or habilitation professionals. The majority of CP research and clinical activities support the use of interventions to maximize each individual's physical well-being and functional skills, and thereby their potential to live life to its fullest.

EPIDEMIOLOGY AND PREVENTION EFFORTS

Much of the information regarding the prevalence and trends in CP comes from population databases in Sweden, Western Australia, and England. Over a period of 40 years, these databases show the prevalence rate of CP to be between 1.5 and 2.5 per 1000 live births (Hagberg, Hagberg, Beckung, & Uvebrant, 2001; Pharoah, Cooke, Cooke, & Rosenbloom, 1990; Stanley, Blair, & Alberman, 2000). Data from the United States are consistent with these studies showing the prevalence of CP at 2.4 per 1000 live births (Boyle et al., 1991). Trend analysis over a 16-year period shows no significant change in the overall prevalence of CP per 1000 live births and 1-year survivors between 1975 and 1991 (Winter, Autry, Yeargin-Allsopp, & Boyle, 2002). This remains consistent despite the fact that neonatal intensive care lowers the risk of mortality among low birth weight infants. A recent report of infants born at 500–1500 g showed that despite a declining mortality rate in this group of low birth weight infants born from 1979 through 1994, the rate of CP did not increase (O'Shea, Preisser, Klinepeter, & Dillard, 1998).

Preventing CP is a large and complex task because there are multiple etiologies. Much attention has been paid to the prevention of premature birth as more than half of children with CP are born at low birth weight or less than 2500 g. Recent information regarding the successful use of progesterone injections to prevent premature labor in women who had already had one premature delivery is promising (Meis et al., 2003). Theories regarding uterine irritability leading to premature delivery with the presence of subclinical or overt infection have led to many trials using antibiotics in pregnant women. These studies have shown benefit to the mother and infant in preventing postnatal morbidity but have not yet been shown to decrease the prevalence of CP. Specific interventions for the premature infant that may reduce CP, such as the use of surfactant that has increased survival and has prevented chronic lung disease, have yet to demonstrate an effect on the overall or birth weight specific prevalence of CP (Soll & Morely, 2001).

CLASSIFICATION OF CEREBRAL PALSY

The classification system for CP serves to highlight consistent differences in the types of CP, the associated conditions, and similarities in etiology. CP is most often congenital, meaning that the cause is presumed to be prenatal or perinatal (with perinatal defined as the time period beginning at the initiation of labor and ending at 30 days postdelivery). CP is less often acquired. Examples of acquired CP include a postnatal infection, trauma, and shaken baby syndrome. CP is classified in two ways. Attempts have been made to classify CP in degrees of severity such as mild, moderate, and severe. The use of this terminology is subjective and is not universally accepted. Most often, CP is described by the terms extrapyramidal and spastic types. Within the spastic types, CP is described by location of the affected area. About 75% of children with CP have spasticity with spastic hemiplegia, diplegia, and quadriplegia being evenly distributed amongst all children with spastic CP.

Extrapyramidal versus Spastic CP. The descriptions extrapyramidal versus spastic refer to the location of the abnormality or damage to the brain. Movement is initiated and regulated by the motor control system of the brain. The pyramidal tracts carry the signal for muscle contraction. The extrapyramidal system, including the basal ganglia, provides regulatory influences on that contraction.

Extrapyramidal CP is a term used to describe the abnormal movement patterns that are caused by damage to areas of the brain outside of the pyramidal tracts. Extrapyramidal CP is often further subdivided into dyskinetic and ataxic types. Dyskinetic movement disorders within CP classification systems include dystonia, athetosis, chorea, rigidity, and hypotonia. Often, more than one of these movement patterns are present.

Choreoathetoid CP, an example of extrapyramidal CP, occurs when the damage to the brain is located in the basal ganglia, classically kernicterus (Sugama, 2001) (Centers for Disease Control and Prevention, 2001). The brief, jerking, purposeless movements of chorea and the long axial writhing movements of athetosis characterize choreoathetoid movements.

Dystonic CP is characterized by the simultaneous involuntary contraction of agonist and antagonist muscle group. This pattern is often fluctuating.

Hypotonic CP is a term that creates confusion. Hypotonia is described as low resting muscle tone. When hypotonia is generalized throughout the body, the term CP is often applied. In these cases, it impairs function, is nonprogressive, is a result of damage to the developing brain, and is not due to a known other cause. When this low muscle tone accompanies a condition like Down Syndrome or Prader–Willi Syndrome, the term hypotonic CP is not added to the diagnosis but hypotonia is understood to be an aspect of the underlying primary, genetic condition.

Ataxic CP, a subtype of extrapyramidal CP, results when damage occurs in the cerebellum. Ataxia is characterized by difficulties performing

finely controlled movements and results in overshooting when reaching for objects and a wide-based, unsteady gait.

Spastic CP is a movement disorder characterized by velocity-dependent increase in resistance. Approximately 75% of cases are primarily *Spastic CP,* which is a movement disorder characterized by velocity-dependent increase in resistance. Spasticity results when damage occurs along the pyramidal tract. Attempts to localize the spasticity and describe spastic CP in terms of where on the body it is expressed aid understanding; in general, of the etiology, functional impairment, associated conditions, and potential treatments for children with spastic CP.

Diplegic CP describes spasticity that occurs in the lower extremities. An individual with diplegic CP, however, will often experience coordination difficulties of the upper extremities or even spasticity of the upper extremities. But, the predominant impairment is in the lower extremities.

Hemiplegic CP is when the spasticity is on one side of the body. Typically, the arm is more affected than the leg and may assume a triple flexion posture. The term triple flexion posture describes the classic position of the involved upper extremity with flexion of the wrist and elbow, and adduction of the shoulder.

Quadriplegic CP involves all four extremities. Generally, the legs are more involved than the arms. Individuals with quadriplegic CP frequently present with truncal hypotonia and extremity hypertonicity.

Monoplegic CP is a term used to describe spasticity found in only one extremity. Practically, this situation is likely the result of a hemiplegia in which the impairment of the leg is imperceptible.

As with any classification system, the present nomenclature does not completely describe what is seen in practice. Many individuals with CP have a combination of the previously described movement abnormalities. This is referred to *as mixed pattern CP* and is present in many individuals with CP. Because epidemiologic studies often use a primary diagnosis for classification, mixed pattern CP is likely to be underreported. Additionally, as children grow and develop, the pattern of CP, and specific diagnosis, may change.

DIAGNOSING CEREBRAL PALSY

Many types of physicians are called upon to make this diagnosis including primary pediatricians, family practitioners, developmental pediatricians, pediatric neurologists, rehabilitation specialists, pediatric orthopedists, and geneticists. Diagnosing and imparting the diagnosis requires an understanding of the risk factors for CP, experience with the neurologic examination that is unique to the developing child, use of imaging studies, and the ability to give the diagnosis to the family and child in a compassionate, realistic, but hopeful manner.

In reviewing the patient history, it is helpful to gather information about factors that are related to CP as an outcome. Oftentimes, it is easiest to consider these risk factors in three broad categories as, prenatal,

perinatal, and postnatal risk factors. Infants born with intrauterine growth retardation are at risk for a variety of developmental disabilities including CP. Growth retardation may be a result of an injury or developmental abnormality of the brain or a genetic defect. Recently more genetic disorders have been identified as causes for CP. Congenital malformations of the brain are most often idiopathic, but a few of these disorders have been found to have specific chromosomal abnormalities, such as the deletion in the short arm of chromosome 17 in the Miller–Dieker form of lissencephaly (Dobyns et al., 1996). Stroke or hemorrhage occurs in the prenatal period. Occasionally the cause of such an event is known, such as maternal anticoagulation therapy resulting in fetal CNS hemorrhage or Factor V Leiden mutation, which causes resistance to activated protein C, causing fetal stroke (Thorarensen, Ryan, Hunter, & Younkin, 1997). Often, evidence for the stroke is found on head imaging studies without a specific reason being evident. The presence of calcifications on a head computer tomography (CT) may suggest an intrauterine infection such as cytomegalovirus or toxoplasmosis.

Thorough investigation into the circumstances of the child's birth is important. The neonatal discharge summary should be scanned for information on birth weight and gestational age, Apgar scores, head ultrasound results, and presence of neonatal seizures. It is known that the risk of CP increases with decreasing birth weight. Nevertheless, even in the majority of children born at extremely low birth (<1000 g) only 17%, showed signs of CP at age 18 months in one recent study (Vohr et al., 2000). In premature infants, the presence of an intraventricular hemorrhage and parenchyma involvement on head ultrasound studies is predictive of CP (Msall et al., 1994; Pinto-Martin et al., 1995; Vohr et al., 1999).

Although the prevalence of neonatal asphyxia is low, it is devastating. The cause of the asphyxia may be known, such as prolonged hypoxia (in the event of a placenta abruption). In other cases, its onset is less defined but the neonate was clearly affected as evidenced by severely depressed Apgar scores and or neonatal seizures within the first 24 hr of life. Postnatal causes for CP tend to be more obvious and dramatic such as a shaking injury, accidental trauma, and bacterial meningitis.

Diagnosis of CP

The diagnosis of CP consists of specific neurologic exam findings present in the developing child and a history that confirms the motor delay or deviance is not deteriorating. The neurologic findings include the following: (1) abnormality of muscular tone, (2) abnormalities of posture/movement, and (3) persistence of primitive reflexes. Findings consistent with central nervous system injury or congenital brain anomaly on head imaging studies are supportive but not diagnostic of CP.

Motor delay in a child is most notable when a child fails to accomplish known gross motor milestones. Fine motor milestones receive less attention by both parents and medical professionals but can signal the presence of CP. Normal gross and fine motor milestones are listed in Table 3.1.

Table 3.1. Motor Milestones in Normal Child Development

Gross motor milestone	Age	Fine motor milestone	Age
Head off table	1 month	Retain rattle	1 month
Chest up	2 months	Hands unfisted	3–4 months
Roll	3–5 months	Transfers objects	5 months
Sitting without support	7 months	Immature pincer grasp	7–8 months
Cruising	9–10 months		
Walking alone	12 months	Release	12 months
Jump in place	24 months	Hand preference	18 months
Pedal tricycle	30 months		

Walking is the most recognized developmental milestone and occurs in most children at 12 months with the upper limit of the normal range being at 16 months. The presence of persistent head lag, poor sitting balance, inability to bear weight on the legs, and the inability to walk by the appropriate time are other common triggers for an evaluation. Hand preference should not occur prior to 18 months. Hand preference at 6–12 months is often misinterpreted by parents as early hand preference rather than an abnormality of neurologic function in the nondominant hand. This is a frequent presentation of hemiplegic CP and should prompt a neurologic evaluation.

Assessing muscular tone in a child is a skill that requires experience and exposure to children and adults with abnormal tone. Spasticity is the clasp knife response or velocity-dependent increased resistance to passive stretch. When severe, it is an obvious finding. It is often accompanied by increased deep tendon reflexes. Spasticity is more pronounced when a child is anxious or ill. Hypotonia, or low resting muscle tone, can also be present to a greater or lesser degree. It can often be elicited in the smaller child by noticing slip through when holding a child under the arms in vertical suspension. Deep tendon reflexes are either decreased or increased with hypotonia. Rigidity is classically described as lead pipe hypertonia, a persistence of increased tone throughout the range of motion about the joint. Children with rigidity tend to have more fluctuation in tone and have persistent primitive reflexes. Muscular tone can change as a child develops. This is commonly seen in infants with hypotonia who evolve to have spasticity by 12–18 months of age. This change is not due to a progression in the neuronal injury or abnormality but rather reflects maturation of the central nervous system.

The diagnosis of CP requires the examiner to observe movements and postural control that are not typically noted in a neurologic exam. A neurodevelopmental exam of the gross and fine motor systems must be added. Abnormal movements should be noted. Unless specific attention is paid to how a child moves, abnormal movements and postures will be missed on the typical neurologic exam such as combat crawling, dragging one side, or standing on toes. Abnormal postural control is often present in children with CP. The infant exam should assess the quality of head control, rolling, sitting balance, crawling, cruising, and walking. This exam will

pick up abnormalities even if the motor developmental milestones are intact. Attention to the fine motor exam will permit the examiner to detect the persistence of fisting or the presence of ataxic hand movements long before delays in walking are noticed.

Knowledge of primitive reflex patterns is essential. The newborn child has primitive reflexes that are normal and diminish over a period of approximately 6 months. These reflexes must disappear to allow a child to attain normal postural and righting responses acquired from approximately 3–9 months. With CP, this normal evolution is disrupted. The persistence of a startle response or Moro reflex prohibits good sitting balance. The presence of a plantar grasp impedes an infant's ability to cruise. These abnormal patterns will be noticed when specific attention is paid to the neurodevelopmental exam.

Brain Imaging

As previously mentioned, imaging studies can be supportive to but are not diagnostic of CP. Significant changes are occurring in the ability to image the central nervous system. Magnetic resonance imaging (MRI) is the single most useful tool to enhance the history and physical exam in the diagnosis of CP. In children with a major motor delay, the majority will have abnormalities on MRI (Candy, Hoon, Capute, & Bryan, 1993). Head ultrasound is limited to the neonate and young infant while the fontanels remain open. Since MRI is superior to CT in differentiating soft tissue differences such as gray and white matter, the MRI is the recommended study beyond infancy. Positron emission tomographic (PET) scans, functional MRI, and diffusion-weighted imaging may add to our understanding in the future. Currently they are not used as a clinical tool (Chugani, 1993; Hoon & Melhem, 2000; Inder et al., 1999). Magnetic resonance spectroscopy, the observation of intracellular cerebral metabolites, is being used as an adjunct to the information provided by the MRI.

Giving the Diagnosis

For most parents, the moment they hear the diagnosis of CP applied to their child, is a profound event. The diagnosis should be given in terms that can be understood by the parents. Information should be given in written form to provide support to what was discussed in the clinical setting. References for parent/caregiver focused information are included in Appendix A. A follow-up visit to review this information is recommended.

There are certain factors that must be appreciated surrounding the diagnosis of CP. Sometimes, the diagnosis of CP is the least of a parent's concern. Describing CP and its functional implications, its causes, and its treatment may be secondary in the minds of parents who are facing a diagnosis of mental retardation or autism in addition to CP. Hearing that a child has CP and may limp when it walks may be good news to a family faced with the possible death of their child from extreme prematurity. The clinician must try to understand how the diagnosis fits in the context of

what the parent has experienced up to the moment of diagnosis, which will help when trying to gauge the language and terminology that will be most helpful to a parent's understanding.

INTERVENTIONS: A MULTIDISCIPLINARY PROCESS

Facilitating the growth, development, and ongoing progress into adulthood of people with CP requires an integration of family, medical, educational, therapeutic, and technologic support systems. Most major urban areas have a multispecialty clinic that serves many of the needs of individuals with CP and has information and connections to other CP-specific services. Usually these clinics have nursing, orthopedics, developmental pediatrics, psychiatry, physical therapy consultation, and orthotic vendors. Many clinics also have psychology, neurology, occupational therapy, social work, and nutrition as well as close working relationships with many other subspecialists.

While every person presents with a unique set of circumstances, there are common conditions that occur within each type of CP. Broad generalizations about associated conditions can be made to help organize proactive planning for assessment of needs and services for individuals with CP. Table 3.2 shows frequently associated conditions occurring with the common subtypes of spastic CP. As a group, these conditions are associated with these types of CP. In any individual person, all or none of these conditions may exist.

Cognitive Impairment and Learning Disability

Overall, approximately 50% of individuals with CP have some degree of cognitive impairment (Murphy, Yeargin-Alsopp, Decoufle, & Drews, 1993).

Table 3.2. Conditions that are Associated with Specific Types

Hemiplegic cerebral palsy	Spastic diplegia	Spastic quadriplegia
General health good	General health good	Multiple medical conditions: Constipation
Unilateral spasticity	Spasticity of the lower extremities	
Unilateral muscle contractures		Oromotor dysfunction
	Muscle contractures lower extremities	Aspiration and/or pneumonia
Unilateral growth disturbances		
	Learning disabilities	Failure to thrive
Epilepsy	Strabismus	Epilepsy
Visual disturbance: Hemianopsia		Orthopedic problems: Muscle contractures
Learning disability		Hip dislocation
		Spine curvature
		Cognitive impairment of CP

Often, CP is erroneously linked with intellectual disability as if the two conditions always occur together. In addition to intellectual disability, another 25–30% has specific learning disabilities. Evaluation of these concerns is conducted by standardized intelligence and achievement testing usually performed by psychologists. Specialized testing materials, procedures, and interpretation may be required for individuals with significant motor impairment and oromotor dysfunction. Physicians, psychologists, and educators should be vigilant for these learning problems and proactively initiate testing early in the child's educational course so that the findings can be translated to the best individualized education program for the child and individualized services for adults.

Role of Schools

Many advances in the education of children with CP have occurred in the last 30 years. Most of them are the direct result of Individuals with Disabilities Education Act (IDEA) of 1975 also known as Public Law-142. Under this legislation and its amendments, standards are set and some financial support is provided to direct states to provide free and appropriate education to all children with disabilities. The fundamental features of these laws include inclusive education, early intervention, assistive technology, and transition planning. Children with CP qualify for early intervention services based on developmental delay judged by defined criteria. Usually children with CP qualify based on motor delay but may also qualify in other domains. Services to the infant and toddler can be provided in a home-based model or a center-based model. A transition plan is then provided through the individualized family service plan (IFSP) for a preschool program. Some children with CP have no educational needs at this stage but need related services, usually physical therapy. This can be arranged. However, many children will need educational services that will be provided based on the individualized education program (IEP). The IEP will be reviewed and updated periodically throughout the child's educational years. Children with CP often need services such as, physical, occupational, and speech and language therapy. All children with CP will need adaptive physical education, even the child with the minimal functional limitations. In 1991, as a part of PL-102-199, children with CP and other disabilities were made eligible for technology-based assistive devices for educational purposes. Devices commonly used by children with CP include augmentative communication devices, powered mobility, ambulation devices, computers, prescription lenses, and environmental controls. School systems can vary greatly with respect to the extent to which they acquire and utilize assistive technology. Part of transition planning services and providing inclusive education is the ability of the child with CP to move about in the educational environment. Accessible classrooms, bathrooms, and playgrounds are essential to the education of children with these physical disabilities. It is the responsibility of the clinical team assisting the family to advise them about needed services, environmental modifications, and assistive devices, and to advocate with the family as necessary.

Another feature of transition planning is the transition out of the secondary education into higher education, vocational training, and supported employment and other adult services. This planning should and typically does begin years before the end of high school. For the student who is college bound, accessibility should be addressed prior to enrollment. States provide through lead state agencies for vocational rehabilitation services, programs to support vocational training to persons with disabilities. These programs are made available by federal funds dispersed to states meeting the standards set by law. These programs are unlike IDEA, in that this legislation is not enforceable by rights and procedures, as entitlements. However, many private foundations, commercial businesses, and community agencies provide opportunities for persons with CP to find supported employment and alternative day involvements and services.

Social Adjustment

Children with CP may go through a pattern of adjustment to their disability that is fairly predictable depending on their cognitive abilities. Typically, the toddler or preschooler does not notice the difference between them and other children. When the child approaches the later years of preschool and certainly by elementary school, the child with CP realizes these differences and may experience frustration, sadness, and anger at their limitations. Fortunately, other children who are able-bodied are very accepting of differences at this stage. As the child approaches middle school, prejudices against persons with disabilities may develop. Cognitive immaturity or a disruption of what is expected in a peer may account for the negative attitudes about persons with disabilities (Harper, 1999). Hopefully, the child with CP has strong family support to validate their worth as a person and value to their family. Family, educators, and medical professionals can limit the lack of knowledge that can stimulate cruel comments or treatment of the child with CP. This can be done by honest and appropriate sharing of information about CP and education in the schools and in the community.

Adolescence is a turbulent time for all teens, including those with CP. Extended periods of normalizing activities, like clubs, church, music and sports lessons, and opportunities for independence help children with CP accept their differences. Counseling may be necessary at any of these stages of adjustment, and may include a focus on alleviation of distress or teaching of coping and social skills.

MEDICAL MANAGEMENT

Every individual with CP should have a primary physician and medical home. The complexity of medical care is more easily addressed with the assistance of someone who is charged with overseeing the medical care. While this person need not be an expert in CP, an interest in the individual,

a willingness to learn, and expertise in general medical healthcare are necessary to provide an optimal medical home.

Growth

Unlike other children in the United States, those with CP often struggle to maintain or gain weight and are rarely overweight. Many factors contribute to the common problem of failure to thrive that is usually described as weight for height that is less than the 5 percentile on the growth chart (Hamill et al., 1979). There are limitations to the growth chart. Experts in the field of CP and growth recommend the use of skin fold thickness as a measure of nutritional status, because it is a more sensitive measure of failure to thrive (Samson-Fang & Stevenson, 2000).

Most often the cause of failure to thrive in children with CP is organic. Weight gain is based on consuming a larger number of calories than are expended. Children with CP have problems with both energy intake and energy expenditure. A child may be in constant motion or use a large amount of energy to move expending a large number of kilocalories daily. In addition, problems with oromotor function may cause gagging, choking, and/or aspiration of food into the lungs. These factors inhibit ingestion of adequate calories. Oftentimes, a combination of multiple factors such as oromotor dysfunction, muscle spasms with pain, poor sleep, constipation, and behavioral refusal conspire to prevent adequate nutrition and subsequent growth. Addressing all of the factors contributing to poor growth in the individual case will be more effective than concentrating on one single factor. Ultimately, a gastrostomy tube may be a necessary intervention on either a temporary or permanent basis. Parents will often express a sense of relief once a gastrostomy tube is placed.

Oromotor Dysfunction

Oromotor dysfunction is often present in individuals with spastic quadriplegia and children with extrapyramidal CP. Language delay may indicate cognitive delays or may instead be a sign of oromotor dysfunction. Expressive language delay with relatively higher receptive language skills may indicate significant oromotor dysfunction. Children with oromotor dysfunction often benefit from assistive communication devices but need professional expertise to teach a child how to use the device (Downey & Hurtig, 2003).

Drooling is another manifestation of poor oromotor control. Drooling may be worsened by poor head and neck control and decreased sensitivity to saliva in the mouth. The presence of drooling can be very socially limiting for the individual with CP. Multiple interventions are possible for this problem including therapy, medications, and surgery (Blasco & Allaire, 1992).

Oromotor dysfunction may result in aspiration of food or saliva into the lungs. The symptoms of aspiration may be obvious such as choking and/or cyanosis with eating. Aspiration may also occur silently, causing

no discomfort to the individual at all. In this case, a child may experience failure to thrive, frequent pneumonia, or recurrent wheezing. Chronic aspiration should be considered when CP, oromotor dysfunction, and recurrent wheezing are present.

Gastrointestinal Dysmotility

Problems with delayed gastric emptying, gastroesophageal reflux, and constipation are frequent in individuals with spastic quadriplegic CP. These disorders are interrelated and compound one another. The child who has delayed gastric emptying is likely to reflux from the stomach causing pain and discomfort. Thus, eating becomes more difficult resulting in decreased intake of food and fluids causing constipation that, in turn, causes bloating, distension of the abdomen, and poor gastric emptying. This is a problem most often seen in children who have poor mobility and poor nutritional status. Improving nutritional status and hydration through dietary supplements, feeding therapy, and or a gastrostomy tube can break this cycle. Increasing movement and exercise can also improve gastrointestinal dysmotility.

Spasticity Management

Treatment to minimize spasticity does not address the primary pathology of CP that resides in the brain, but it may decrease the sequelae of the increased tone. Problems such as musculoskeletal deformity, limitations of movement, and pain may be minimized by the use of interventions to reduce tone. Historically, diazepam and related medications were the only options for management of spasticity. Within the past two decades, selective medications and surgical interventions have become available. Treatment options for spasticity currently include oral medications, injection of medication into the affected muscle, intrathecal medication, and neurosurgical intervention.

Oral Medications

Common oral medications used to decrease spasticity include diazepam, baclofen, and dantrolene. Diazepam (Valium™) is readily available, inexpensive, and has a long history of use. Unfortunately, diazepam acts centrally and sedating side effects usually make this drug an undesirable choice for managing the problems of spasticity (Young, Robert, & Delwaide, 1981).

Baclofen (Lioresal™) acts at the spinal cord stimulating gamma aminobutyric acid (GABA) inhibitory pathways to reduce spasticity. In large doses it may cause central nervous system sedation. Acute withdrawal from the drug may cause serious central nervous system effects such as hallucinations and seizures or acute increase in spasticity. No intravenous preparation of baclofen is available. Therefore, it is recommended to taper use of this drug prior to surgery or procedure that results in an ileus

(Krach, 2001). Dantrolene sodium (Dantrium™) produces muscle relaxation by interfering with the release of calcium from the sarcoplasmic reticulum of the skeletal muscle. Dantrolene does not directly affect the central nervous system but has central side effects. Dantrolene has the potential for fatal and nonfatal liver toxicity and may cause excessive weakness (Gormley, 2001).

Intrathecal Baclofen

Severe spasticity can be controlled by placing baclofen in a hockey puck-sized device in the abdominal cavity, a treatment called continuous intrathecal baclofen. The baclofen is delivered from the reservoir through a catheter into the spinal canal. The tip of the catheter is usually positioned on the low thoracic area. This externally programmable device pumps very small amounts of baclofen on a continuous basis and can be varied throughout the day by the programmable pump. Complications may include mechanical failure, CNS side effects of lethargy and sedation, and operative complications including infection, disconnected tubing, catheter breakage, cerebrospinal fluid (CSF) leak (Albright, 1996). Despite these limitations, many centers report improvements in muscle tone, range of motion, and functional activities in patients receiving the intrathecal baclofen (Butler & Campbell, 2000).

Nerve blocks

Nerve blocks can be used to affect muscle tone in a localized way. Two examples of chemical neurolysis are phenol blocks and botulism toxin A injections. Phenol, alcohol, and localized anesthetics have long been used to provide a local motor point block. Phenol denatures protein in the myelin sheath of the nerve, destroying the axon. Because nerves are capable of regeneration within 3–6 months, this effect is temporary. Botulism toxin acts to irreversibly block the cholinergic receptors of the neuromuscular junction thus inhibiting the release of acetylcholine, which is required for muscle contraction. This too is temporary, in effect, lasting approximately 2–3 months. Botulism toxin can be rapidly administered without the need for electrical stimulation, and requires a much smaller volume and smaller needle for infusion, thereby causing less pain than the process of injecting phenol. The cost of botulism toxin is quite high compared to other neurolytic agents. The efficacy of single or repeated botulism toxin injections on functional abilities in the long run is unknown. However, use of botulism toxin injections has demonstrated short-term improvement in range of motion, muscle tone, and functional abilities (Koman et al., 2001).

Selective Dorsal Rhizotomy

Finally, spasticity may be permanently addressed with the selective dorsal rhizotomy. This procedure involves the selective cutting of sensory

nerves returning from the lower extremities to the lumbosacral spine. Following the surgery, an intensive rehabilitative program is undertaken by the child to "relearn how to walk." Outcome studies of the selective dorsal rhizotomy show that it helps reduce spasticity and has a positive effect of gross motor function (McLaughlin et al., 2002).

ORTHOPEDIC MANAGEMENT

Orthopedic intervention involves managing the sequelae of increased muscular tone that includes treating and preventing skeletal deformity and addressing muscle, ligament, tendon, and surrounding soft tissue contracture. Orthopedic treatment and spasticity management are not mutually exclusive but rather complimentary components of an appropriate medical intervention plan. Surgical intervention to correct deformity or contracture does not equate to normalization of gait pattern or cure of CP. Correction of a deformity of the musculoskeletal system does not eliminate the cause of the deformity. Orthopedic management of children with CP involves complex decisions integrating surgical skills, knowledge about the patterns of CP pathology, natural progression of the different subtypes of CP, and an understanding of child development. This type of surgery is best done by the surgeon who has pediatric expertise and experience in CP. Finally, orthopedic management of CP takes time and consistent follow-up.

Hip

Children with spastic quadriplegia will frequently have problems with growth about the hip due to increased and abnormal balance of tone around the hip joint. Since bones grow according to the forces placed on them, the femoral head and acetabulum may gradually become dysplastic under the influence of spasticity. These forces may cause the hip to dislocate posteriorly and superiorly out of the joint. This is most likely to occur in nonambulatory children with spastic quadriplegia. If the hip is identified as at risk, hip positioning, soft tissue surgery, and osteotomies are considerations.

Foot

The goal of orthopedic intervention on the foot is to preserve a plantigrade (flat on the ground) foot position. This provides the best biomechanical and proprioceptive position for both ambulation and positioning in a wheelchair or stander. Spasticity works against that ideal often pushing the foot into an equinovarus (plantarflexed and medially deviated) position. Bracing with ankle foot orthoses, serial casting, and botulism toxin injections have been effective in treating the plantar flexion contracture. Orthopedic intervention is often necessary and effective when the contracture has become fixed. Gait analysis laboratories can be used to assist in the decision making process (Schwartz, 2004).

Spine

Treatment of spine curvature in individuals with CP is increasingly limited to pediatric orthopedists with a special interest in spinal surgery. Observation, orthoses, and surgical intervention are treatment options. In general terms, bracing is considered at curves measuring approximately 20–25° but less than 40° (Lonstein, Winter, Bradford, & Ogilvie, 1995). Spine braces should be used to hold a curve rather than providing correction. Progression of the curve is likely in children with neuromuscular curves. A decision to operate, which includes fusion of vertebral bodies to prevent progression and sometimes correction of the curve, is complex.

THERAPEUTIC INTERVENTION

The goal of therapy is to allow a child to participate as fully and developmentally appropriate as possible in their lives. This is done in many ways but traditionally this involves initiating physical therapy, occupational therapy, and speech language therapy. Therapy is provided in a variety of models. It can be provided in the context of a home-based model, a center-based program, a hospital outpatient model, and a multidisciplinary educational model. Therapy can be provided in group sessions or as individual therapy. Finally, therapy can be provided as a direct service or in an indirect consultative fashion.

Specific principles of therapy related to CP are beginning to emerge with new research on the efficacy and outcomes of therapeutic intervention. The goal with all children with CP is to establish normal motor development and function and to prevent contractures and deformity. Therapeutic goals include teaching a developmental sequence of acquiring skills and preparing the child with tone abnormalities to perform specific functional tasks in real life settings. Specific techniques are being studied in children with CP. Forced used or constraint-induced therapy is when the affected side is forced to be used by restraining, by tying back, or casting the typical side of the body. This is a therapeutic approach being tried in adults with hemiplegia resulting from a stroke (van der Lee, 1999). It is also being evaluated for efficacy in children with hemiplegic CP (Willis, 2002). Strength training for a person with spasticity is being promoted now rather than discouraged as had been in the past. Evidence exists that spastic muscles that are weak can improve in strength without increasing the spasticity (Dodd, Taylor, & Damiano, 2002). Therapists may guide the family into thinking about occasional episodes of therapy rather than years of commitment to a therapy program as an expected course of treatment.

INTERVENTION AND SUPPORT FOR ADULTS WITH CP

High functioning adults with CP have a life expectancy close to that of the general population. However, life expectancy can be reduced by factors

that impair an individual's functional level such as low cognitive level, involvement of all four extremities, poor mobility, and dependent feeding (Strauss & Shavelle, 2001). As most individuals with CP live a long life, planning for adulthood is important.

Medical Care

Caring for the medical issues of adults is often overlooked. Adults with CP are often concerned about pain, decline in mobility, fatigue, and loss of independence. In one survey of adults with CP, 35% reported decreased walking ability and 9% reported having stopped walking (Andersson & Mattsson, 2001). Eighteen percent of these respondents reported pain everyday. Pain in individuals with CP is likely to come from the hip and in nonambulatory individuals pain is very common. One survey of nonambulatory adults, with a mean age of 27 years, reported that hip pain was present in 47% of respondents. Of that group only 13.6% of those with pain had received medical treatment for the pain (Hodgkinson et al., 2001).

Osteoporosis is a concern for all adults, but especially for those with CP. In fact, osteopenia was found in 77% of a population-based cohort of children and adolescents with CP and 97% of the subgroup of children who were older than 9 years and could not stand (Henderson et al., 2002). Given this knowledge and the known patterns of bone density loss in the general adult population, concern for increased bone fractures, subsequent pain, and functional limitation, more research into treatment is needed.

The medical care and treatment of patients with CP has usually been provided by pediatricians and pediatric subspecialists. The medical needs of the adults with CP may be left untreated, as few medical professionals caring for adults are familiar with CP and its related conditions. Oftentimes, unrelated acute and chronic conditions go untreated as adults with CP find it difficult to find routine medical care and dental care. Routine care also may be limited by financial disincentives to care for individuals with disabilities and physical barriers present in clinic settings.

Vocational Services

Employment for adults with CP and other disabilities, including intellectual disabilities and epilepsy has improved through vocational services. An increasing number of individuals with CP are gaining competitive employment (Gilmore, 2000). Successful employment is partly related to type and severity of disability, intelligence level, functional independence, and whether or not assistive technology is needed (O'Grady, 1995). Community integrated employment must include an appropriate and reliable means of transportation. Driver assessments through the Bureau of Motor Vehicles and handicapped-accessible public transportation are means to overcome obstacles to competitive employment. Unfortunately, despite the increased prevalence of competitive employment, there was a decrease in real earnings when earnings were adjusted for inflation (Gilmore, 2000). Private groups such as United Cerebral Palsy, professional groups such

as the Academy of Cerebral Palsy and Developmental Medicine, and federal agencies such as the Administration of Developmental Disabilities and the Centers for Disease Control and Prevention are trying to address the concerns of these adults. Other advocacy groups also exist to help adults with CP.

SUMMARY

CP, present in approximately 2 per 1000 live births, is a group of motor impairment syndromes caused by an injury or abnormality of the brain that is nonprogressive. CP is classified by two main categories; spastic and extrapyramidal. The diagnosis is made by physical exam and a thorough history. Taken together, the presence of abnormal tone, abnormal movements and postures, and persistence of primitive reflexes in an infant with motor delays, which are nonprogressive, constitutes the diagnosis of CP. MRI is a helpful adjunct but is not diagnostic of CP. Educational services and related services such as physical therapy and assistive technology are available in school systems as a result of the IDEA legislation. Adjusting to having CP is difficult. A strong family that is able to validate the child's worth and contribution to the family will make this process easier.

Most children access the services of a multidisciplinary CP clinic to address the many associated conditions and needs related to CP. These issues include but are not limited to initial diagnosis, musculoskeletal problems, spasticity management, feeding difficulties, weight problems, respiratory infections and aspiration, epilepsy, and impaired communication, learning, and mobility. Relatively new treatments for spasticity include selective dorsal rhizotomy, intrathecal baclofen pump placement, and botulism toxin injection. Research has demonstrated that spastic muscles do benefit by strengthening despite earlier concerns that strengthening would only result in increasing spasticity.

Adults with CP are concerned about persisting or chronic pain, osteoporosis, and inadequate primary medical care due to misunderstandings about CP, paucity of providers available to care for their specialized needs, and financial barriers to providing care.

APPENDIX A

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