Cerebral Palsy: Prevalence, Predictability, and Parental Counseling

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Abstract

Cerebral palsy (CP) is the most common cause of severe physical disability in childhood, occurring in approximately 2 in 1,000 liveborn infants. Although the prevalence of CP appears to have stabilized in the past 2 decades, recent studies suggest that severe CP may be decreasing. Neuroimaging studies help identify abnormal neuroanatomic findings, which are found in most affected children. Neuropathology identified by magnetic resonance imaging (MRI) corresponds well to clinical descriptions of motor impairment in children who have CP. Clinical risk factors, combined with imaging studies, can help identify a subpopulation of infants who are at high risk for poor neurodevelopmental outcome. Counseling caregivers on future adverse developmental risks can be challenging for the clinician in the neonatal intensive care unit (NICU), especially because the cause of CP remains unexplained in most cases and is typically diagnosed outside the neonatal period. Early counseling of families of at-risk neonates may function as the starting point for parental adaptation to a lifelong condition that requires ongoing services and adjustments to promote the overall health and well-being of their child.

Objectives After completing this article, readers should be able to:

1. Define cerebral palsy (CP).
2. Discuss the epidemiology and risk factors for CP.
3. Address the capabilities of neuroimaging to predict and classify CP.
4. Review the neonatologist’s role in discussing and disclosing information to families related to long-term outcome in infants at risk for CP.

Introduction

CP is the most common cause of severe physical disability in childhood. (1) This lifelong condition may alter the trajectory of many aspects of a child’s development, both primarily and secondarily. In the United States, it is estimated that approximately 764,000 children and adults manifest one or more of the symptoms of CP and that 10,000 babies born annually develop CP (United Cerebral Palsy, http://www.ucp.org/uploads/media_items/cerebral-palsy-fact-sheet.original.pdf, accessed March 2011). Loss of productivity, dependency, progressive deterioration of motor physical function, recurrent use of rehabilitation services, and reduced life expectancy all contribute to the economic burden, which has been reported in different global settings. (2) The Centers for Disease Control and Prevention has estimated the average lifetime costs per person who has CP to be $921,000 (in 2003 dollars), with lifetime costs for all affected people who were born in 2000 to be $11.5 billion. (3) The Danish Cerebral Palsy Register, which has registered about 50% of the Danish population of individuals who have CP since 1950, estimated the lifetime cost of CP at about €860,000 for men and about €800,000 for women ($1,317,200 and $1,225,600, respectively, for 2008 exchange rates).

Definition

CP is an umbrella term describing multiple diseases originating early in life that are characterized by variable motor impairments due to unspecified causes and cerebral pathologies. No definitive diagnostic tests are available. The current definition of CP was

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published in 2005, based on the conclusions of an International Workshop consisting of experts in the field of developmental disorders:

“Cerebral palsy describes a group of disorders of the development of movement and posture, causing activity limitation, that are attributed to nonprogressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, cognition, communication, perception, and/or behavior, and/or by a seizure disorder.” (4)

The definition excludes progressive brain disorders as well as neurodevelopmental disabilities lacking aberrations of movement and posture. Although the brain pathology in patients who have CP is static, motor ability impairments related to CP may change over time. Various secondary conditions that are not a part of the primary disabling condition but occur as a result of the condition may adversely affect function and quality of life. (5)

CP should not be viewed as a singular diagnosis because the term CP encompasses multiple etiologic diagnoses. It is best considered a descriptive label based on a broad range of presentations that include type, severity, and bodily distribution of primary motor impairment, associated nonmotor neurologic and behavioral impairments, functional deficits, and cerebral pathology. A similar clinical pattern of CP may result despite different causes at different developmental stages; alternatively, a similar cause may produce variable clinical patterns of CP. Thus, clinical classification provides insufficient insight into the cause of CP. Although the CP label does not specify a particular cause or pathology, affected children are typically grouped into phenotypic subtypes based on the distribution of limb weakness and type of tone abnormality. CP is categorized as spastic, ataxic, or dyskinetic, and the distribution is categorized as bilateral or unilateral. Most cases of CP among children born preterm are of the spastic type, rather than the athetotic or dyskinetic types. (6) The most common spastic subtypes in these children are diplegic (bilateral limb involvement) or hemiplegic (unilateral limb involvement). The diagnosis of CP is typically made after the age of 2 years, but identification of the patterns of aberrant motor posture and function associated with CP can be made as early as 6 months of age. (7)

**Prevalence**

Approximately 2 in 1,000 liveborn children suffer from CP. (8)(9) Globally, the reported incidence and prevalence of CP varies by region, population, age, and severity, which may limit the generalizability of population-based results. Determining who to include in population-based CP registers or surveillance systems remains a challenge because of the lack of universal agreement on age cutoffs for determining nonprogression of CP.

Tracking trends of gestational age-specific CP prevalence along with gestational age-specific survival is important to clarify if improved survival is linked to increased prevalence of CP. Because neonatal intensive care is dynamic, survival and outcome trends should be evaluated periodically to assess the effectiveness and sustained improvement in care practices. Focusing on peak CP prevalence rates in population-based studies conducted over long time intervals may misrepresent recent trends reflecting decreased CP prevalence associated with improved survival. The true prevalence of CP may be underreported because severely impaired infants may die before developing findings that are diagnostic of CP or before their abnormalities meet criteria for a diagnosis of CP. In addition, if children who have mild CP are not brought in by their parents for evaluation, the true prevalence of CP in population studies is underreported.

**Prevalence in Term Infants**

The risk of CP among term infants is much lower than in preterm infants, although most CP is associated with term deliveries because most infants are born at term. Globally, CP prevalence data show some geographic differences, but overall, population-based reports have shown a fairly stable rate among the term group at 1 to 1.5 per 1,000 live births. (10)(11)(12)(13)(14) Although the overall prevalence of CP has been stable, recent studies suggest that severe CP may be decreasing, as demonstrated by studies of children who had CP born at term from Iceland and Sweden showing a significant decrease in the proportion that had two or more associated impairments. (14)(15) Affected Icelandic children born at term from 1997 to 2003 had better gross motor abilities, were less likely to suffer from epilepsy, and were more likely to have the diplegic subtype of CP when compared with those born from 1990 to 1996. (14) Data from the Surveillance of Cerebral Palsy in Europe network, a population-based CP register representing the largest international collaboration of CP registers in the world, demonstrated significant changes in the prevalence of spastic CP subtypes in children born with birthweights of at least 2,500 g born between 1980 and 1998. (8) The bilateral spastic form increased from 0.58 (95% confidence interval [CI], 0.41 to 0.80) in 1980 to 0.33 (CI, 0.22 to 0.46) in 1998, and the unilateral...
spastic form increased from 0.37 (CI, 0.23 to 0.58) to 0.46 (CI, 0.34 to 0.62), with a concurrent reduction in neonatal mortality from 1.7 (CI, 1.4 to 2.1) to 0.9 (CI, 0.7 to 1.1) per 1,000 live births. Similar decreasing trends in the proportion of infants weighing more than 2,500 g who had spastic diplegia (62% in 1988 to 1993 and 44% in 1994 to 1998) have been reported in Slovenia. (16) Additional population-based research is needed to determine if functional status and severity of associated comorbid conditions among affected children of various birthweights and gestational ages are also improving over time.

The risk for CP in term deliveries may vary, depending on the timing of birth within the term period. Moster and associates (17) reported a prevalence of CP of 1.15 per 1,000 births (CI, 1.10 to 1.20) based on a population-based follow-up study using the Medical Birth Registry of Norway. CP was diagnosed in 1,938 of 1,682,441 singleton children born between 1967 and 2001 whose gestational ages were 37 through 44 weeks. Delivery at 40 weeks’ gestation was associated with the lowest risk of CP (0.99 per 1,000; 95% CI, 0.90 to 1.08) compared with delivery at 37, 38, or later than 41 weeks’ gestation, which was associated with an increased risk of CP. Whether the timing of delivery (too early or too late between 37 and 44 weeks) increases the risk of CP or if fetuses predisposed to CP have a disturbance in the timing of their delivery is unclear.

**Prevalence in Very Low-birthweight Infants**

Gestational age and birthweight are interrelated factors that both affect CP risk. (14) In a large national Norwegian registry study that included 903,402 liveborn infants, those born between 23 and 27 weeks’ gestation were 78 times more likely to have CP than those born at term. (18) Although children born before 32 weeks’ gestation have an increased prevalence of CP (up to 10%), they contribute to less than 2% of neonatal survivors and to a minority (approximately 20% to 25%) of all CP in developed countries. (19) A prospective population-based North of England Collaborative Cerebral Palsy Survey demonstrated a prevalence of CP in singletons of 8.9 (72/8,082) per 1,000 neonatal survivors whose birthweights were between 1,500 and 2,499 g based on 5-year cohort data from 1996 to 2000. (20) Reported CP prevalence rates vary from 19 to 152 per 1,000 live births for very preterm and very low-birthweight (VLBW) infants. (9)(21)(22)(23)(24) This broad range reflects differences in regional infant mortality rates, which influence the prevalence of CP in the surviving population, as well as sample size differences and changes in practice over time. A limitation of currently available population studies is that the reported CP prevalence rates are often derived from patient cohorts that include pooled data spanning the past 3 decades, a time period during which neonatology care practices have dramatically changed, thus diminishing the utility of this information for parental counseling.

Several epidemiologic studies have shown that as survival of extremely preterm infants has increased, the prevalence of CP initially increased, then stabilized, and subsequently declined. (23)(25) For example, in Northern Alberta, 2,318 infants of 20 to 27 weeks’ gestational age whose birthweights were 500 to 1,249 g were live born from 1974 to 2003. (24) Overall, CP prevalence rates peaked in 1992 to 1994 at 131 per 1,000 live births, decreasing to 19 per 1,000 live births in 2001 to 2003. From 1992 to 1994 and 2001 to 2003, population-based survival increased in VLBW infants from 4% to 31% (P<0.001) for infants born at 20 to 25 weeks gestational age and from 23% to approximately 75% (P<0.001) for infants born at 26 to 27 weeks gestational age. As survival of VLBW infants increased, the prevalence of CP decreased. From 1992 to 1994 and 2001 to 2003, the CP rate decreased from 110 to 22 per 1,000 live births for infants born between 20 and 25 weeks gestational age and from 155 to 16 per 1,000 live births for infants born between 26 and 27 weeks gestational age. (24) These fluctuations likely correspond with advances in obstetric and neonatal care that improve survival of very preterm infants. As new gestational survival boundaries are crossed, the gestation-specific prevalence of CP tends to increase, then decline as neonatal management techniques are refined. (19)

**Neuroimaging**

Thorough understanding of the pathogenesis, causes, and timing of different CP subtypes and severities remains elusive because CP is a complex heterogeneous disorder diagnosed on clinical manifestations. To address this shortcoming, neuroimaging is currently recommended as a standard evaluation for children who have CP. (26) Abnormal neuroanatomic findings are found in 80% to 90% of children who have CP and are detected more often with MRI than with computed tomography (CT) scan, with white matter damage being the most common abnormality seen. (27) Population studies to identify MRI findings in children who have CP may reveal characteristic pathologic neuroimaging patterns associated with specific neurologic subtypes, CP severity levels, and other categorical variables and ultimately serve as a biomarker for future outcomes. (28) Newer...
analysis techniques, such as functional connectivity MRI, volumetric analysis, and surface-based morphometry, are being investigated as tools to provide insight into alterations in structural and functional brain maturation associated with preterm birth. (29)

MRI for Predicting CP

Neuropathology identified by MRI corresponds well with clinical descriptions of motor impairment in children who have CP. MRI findings of unilateral pathology are typically associated with unilateral impairment, periventricular white-matter damage with lower limb spastic impairment (with upper limb involvement increasingly proportional to increasing damage), and basal ganglia damage with dyskinetic impairment. (19) Asymmetric myelination in the posterior limb of the internal capsule on MRI at 40 weeks postmenstrual age is associated with subsequent hemiplegia in infants who have periventricular hemorrhagic infarction. (30) For infants who have periventricular leukomalacia (PVL), bilateral abnormalities in the posterior limb of the internal capsule are associated with spastic diplegia or quadriplegia. (31) Brain malformations such as schizencephaly, lissencephaly, polymicrogyria, and heterotopia have all been associated with spastic CP. These migrational disorders, found more commonly in children who have CP born at term (>37 weeks gestational age) compared with preterm, are characterized by varying levels of abnormal gyral and sulcal development and are more common in children who have hemiplegia, which also occurs more frequently in children born at term. (27)

Abnormal findings on MRI at term-equivalent ages in very preterm infants strongly predict adverse neurodevelopmental outcomes at 2 years of age. (32) The European Cerebral Palsy Study, a cross-sectional population study carried out in eight centers in Europe, demonstrated correlations between MRI findings and types of motor impairments. (33) This study identified 585 children diagnosed with CP after the age of 2 years who were born between 1996 and 1999, of which 431 were clinically assessed and 351 had brain MRI scans. White-matter injury was found in 71.3% of the children who had diplegia (n=87), 34.1% of those who had hemiplegia (n=31), and 35.1% of those who had quadriplegia (n=20). In general, children who had posterior-only or posterior and middle white-matter injury were found to have spastic diplegia, whereas children who had spastic quadriplegia demonstrated damage across posterior, middle, and anterior brain regions per MRI. Most of the preschool-age children who had hemiplegia (89%) and diplegia (69%) were walking compared with only 9% of those who had quadriplegia. Feys and associates (34) evaluated the relationship between neuroradiologic findings (MRI and CT scan) and upper limb function in 53 children evaluated at a mean age of 5 years 7 months (SD 4 y 6 mo) who had hemiplegic CP. Children in the PVL group (brain lesions occurring in the late second and early third trimester) had better upper limb function than those in the congenital cortical-subcortical group (brain lesions occurring in the late third trimester or around the time of birth).

The presence of white-matter lesions on MRI or cranial ultrasonography is a strong predictor of CP. (27)(32)(35)(36) A prospective longitudinal study of 167 very preterm infants who had MRIs at term-equivalent ages and long-term neurodevelopmental follow-up evaluation at 2 years of age corrected for prematurity demonstrated that white-matter abnormalities, especially those that are moderate and severe, were useful markers for the elevated risk of severe cognitive delay, severe psychomotor delay, CP, and neurosensory impairment. (32) However, a substantial proportion of children who had moderate-to-severe white-matter abnormalities were free of severe impairment at 2 years of age. Children who have spastic syndromes frequently have white-matter injury; children who have extrapyramidal syndromes often have basal ganglia abnormalities on imaging. (37)

PVL, a form of white-matter injury, is the most common cause of CP in preterm infants. (38) PVL can result in a cystic necrosis of white-matter tracts or diffuse noncystic lesions, which are now the predominant lesions seen in preterm infants. A strong relationship exists with the finding of cystic PVL and the development of CP. Fewer than 5% of preterm infants in whom repeated ultrasonography shows only increased periventricular echogenicity without cysts subsequently develop overt CP, although substantially more show evidence of cognitive dysfunction. (38) Diffusion tensor imaging has demonstrated significant variability in white-matter injury patterns in patients who have PVL, with the most frequent injury occurring to the retrolenticular part of the internal capsule, posterior thalamic radiation, superior corona radiata, and commissural fibers. (39) PVL may be seen in all subtypes of CP, but it is the primary lesion in patients who have spastic diplegia and likely reflects a cerebral injury that occurred in utero. (33)(40)

Determining the timing of brain injury and whether it was acquired pre- or postnatally remains challenging. Although white-matter injury such as PVL is believed to occur before about 34 weeks of gestation, 25% of the white-matter injury group in the European Cerebral
cerebral palsy

Palsy Study were born at term. (33) In a hospital-based study of 130 children who had spastic CP, Kulak and associates (40) reported that almost 50% the children who had PVL were born at term and had no histories suggestive of perinatal asphyxia or low birthweight. From a Quebec population-based registry (REPACQ), Towsley and colleagues (28) reported that 41 of the 213 children who underwent neuroimaging studies (MRI or CT scan) had evidence of PVL and 15 (37%) were born at term. However, the REPACQ study demonstrated several characteristic differences between term and preterm children affected by both PVL and CP. (41) Spastic hemiplegia was significantly more common in the term group (8/15 [53.3%]) than the preterm group (5/26 [19.2%]), whereas spastic diplegia was more common in the preterm group (9/26 [34.6%]) than the term group (2/15 [13.3%]). Superior motor outcomes were more common in the term group, with 80% (12/15) able to ambulate independently compared with only 46.1% (12/26) of preterm children. These differences reveal that despite a common radiologic pattern, term and preterm children affected by both PVL and CP exhibit clinicopathologic distinctions that may be related to differences in prenatal and perinatal risk factors and the timing of acquired brain injury.

**Limitations of MRI in Identifying and Predicting CP**

MRI and clinical findings do not always correlate well. Normal neuroanatomy is present in 10% to 20% of individuals who have CP, (19)(27) and up to 25% of children who have significant lesions such as PVL noted on MRI do not develop any neurologic disorder. (42) In addition, the location of brain lesions does not always coordinate with the topography of functional disabilities in children who have CP. The bilateral periventricular lesions that are often seen in those who have hemiplegia exemplify this apparent discrepancy. (34) Etiologic interpretations of anatomic image findings are commonly made, despite insufficient evidence to support these associations. Although abnormal MRI findings may correlate with poor neurologic outcomes in children who have CP, physicians should acknowledge the limitations of MRI in accurately predicting the timing of neurologic insult and the severity of long-term neurodevelopmental problems when discussing findings with families of infants at risk for CP. Physicians need to weigh the potential risks associated with sedation and the costs associated with MRI in children who have CP with the benefit of providing families with an improved understanding of neuropathology related to their child’s clinical findings.

**Ultrasonography for Predicting CP**

Craniul ultrasonography has been used extensively in preterm infants to identify maturation-dependent neurologic injury. The presence of more than one abnormal ultrasonographic finding has been associated with an increased risk of developing quadriplegia or more severe forms of CP. (43) Echolucency and ventriculomegaly on ultrasonography are associated with subsequent CP. (32)(35)(43)(44) Ventriculomegaly identified closer to term appears to be more predictive of CP than ventriculomegaly in the first 2 weeks after birth in very preterm infants. (43)(45) Cerebellar injury in term and preterm infants is associated with a high prevalence of long-term pervasive neurodevelopmental disabilities. (46)(47) Messerschmidt and associates (48) studied the role of the cerebellum in neurodevelopment in 31 sex- and gestational age-matched pairs of former preterm infants (mean gestation, 27.0 ± 1.6 wks) followed up at a minimum age of 24 months. Disrupted cerebellar development detected on cranial ultrasonography was associated with impairment of neuromotor and mental developmental outcomes in preterm infants independent from supratentorial brain injury. Mixed (spastic-ataxic, spastic–dyskinetic) CP was diagnosed in 48% of affected patients, whereas none of the patients in the control group had mixed CP.

Specific patterns of white-matter injury (eg, PVL) are associated with increased risks of CP and can be detected using cranial ultrasonography. Himpens and coworkers (49) reported on the predictive value of ultrasonography in 163 (16.1%) of 1,015 children who underwent cranial ultrasonography during infancy in the NICU over an 11-year period (1995 to 2005). CP was noted in 4% of children who had normal ultrasonographic findings and in 30% of children who had abnormal findings. Infants who had PVL grade III or greater were 79 times more likely to develop CP (95% CI, 22 to 282). (49) Intraventricular hemorrhage (IVH), which is seen in preterm infants also at risk for PVL, has been associated with poor neurologic outcomes, including CP. IVH grades III and IV are significant predictors of major neurologic damage, (50) whereas IVH grades I and II detected by cranial ultrasonography are not significant predictors of CP. (51)

**Limitations of Ultrasonography in Identifying and Predicting CP**

Limitations of cranial ultrasonography include poor sensitivity for diffuse white-matter abnormalities detected by MRI, (44)(52)(53) inability to image cortical and cerebellar structures adequately, (54) and dependency on
patent fontanelles to perform imaging that requires a sufficient angle of insonation (anterior fontanelle) to assess the periventricular white matter. (44) Cranial ultrasonography through the mastoid fontanelle can demonstrate injury, such as posterior fossa hemorrhage, missed by using the routine anterior fontanelle approach but confirmed by MRI. (54)

**Neuroimaging and CP in the Low-income Setting**

Data regarding the clinical spectrum of CP in developing countries are limited, and neuroimaging data are even less relevant, despite the majority of births and cases of birth asphyxia occurring in these settings. In low-income countries, birth asphyxia is much more common than in high-income countries, (55) which may influence the subtype distribution in the CP population. Birth asphyxia has been highly associated with CP in developing countries. (56)(57) In north India, Singhi and associates (57) reported a history of birth asphyxia in 71% (25/35) of children who had spastic quadriplegia, with 91.4% (32/35) having abnormal findings on brain MRIs (18/35 with diffuse encephalopathy). Failure to study the epidemiology of CP in low-income settings more thoroughly limits our understanding of CP in areas of the world where most people live. This epidemiologic shortcoming needs to be addressed to appreciate fully the impact and distribution of CP globally. Furthermore, understanding the contributing factors to the CP population can guide global efforts targeting potentially high-yield interventions (eg, training health-care staff in newborn resuscitation and care of low-birthweight babies) to decrease the development or severity of CP worldwide.

**Parental Counseling**

Biomarkers allowing for the accurate prediction of which neonate will develop CP are lacking. Clinical risk factors such as extreme prematurity, IVH, sepsis, or necrotizing enterocolitis, when combined with results of imaging studies, can identify a subpopulation of infants that is at high risk for poor neurodevelopmental outcome. Despite this, a large European study demonstrated that at discharge from the special care infant unit, 47.3% of parents reported that they were unaware of any concern about their child who later developed CP. (33) A challenge for neonatologists is that the diagnosis of CP is not made in the neonatal period but evolves over time. The inherent heterogeneous nature of CP impedes the ability to make an expeditious diagnosis, which may negatively complicate the initial disclosure process of communicating a diagnosis clearly to the family that may have an impact on future family adaptation and well-being. (58) A substantial number of children who eventually are diagnosed with CP come from the NICU environment. Therefore, early counseling of parents of neonates at high risk for CP may function as the starting point for parental adaptation to a lifelong condition that requires ongoing services and adjustments to promote the overall health and well-being of their child. Table 1 lists some potential future morbidities associated with CP. Parents may benefit from proactive informative counseling to educate them about aspects of CP without provoking unnecessary anxiety, particularly given the limitations in definitive prognostic capabilities during the neonatal period for future mobility function. (59) At the time of discharge from the NICU, neonatologists should communicate to pediatricians any concerns for future disabilities in at-risk patients to avoid potential negative affects on parental satisfaction, delayed timing of diagnosis, or inadequate provision of proper resources that ultimately may prevent optimal patient care.

Cranial ultrasonography is commonly obtained in the NICU and may be useful in predicting severe neurodevelopmental abnormalities in preterm infants. (30)(31)(60) However, normal findings neither exclude the possibility that an infant may develop CP nor predict that an infant may develop CP. In the ELGAN study of 1,053 infants born before 28 weeks’ gestation diagnosed with CP at 2 years of age, 43% (51/120) of children who

**Table 1. Morbidities Associated With Cerebral Palsy**

- Cognitive impairments
- Epilepsy: 20% to 40% of patients
- Behavior problems: 5 times more likely in children who have CP
- Pain
- Weakness
- Speech impairment: up to 80% of patients
- Low visual acuity: up to 75% of all children who have CP
- Gastrointestinal and feeding problems: 50% of children who have CP
- Dental caries
- Developmental enamel defects
- Gingival health, tooth wear, oral mucosal health, and malocclusion problems
- Swallowing dysfunctions and dysarthria symptoms
- Stunted growth: 25% of children who have CP
- Under- or overweight problems: 50% of children

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had CP had normal cranial ultrasonography study results. (43) Due to false-negative and false-positive results, clinicians counseling parents based on cranial ultrasonography findings alone should communicate the range of predicted results, including that 1) early abnormalities may resolve; 2) even when abnormalities are persistent, plasticity of the brain may modify the association between ultrasonographic abnormalities and impairments; and 3) an infant whose neonatal ultrasonography results are normal may develop CP or another developmental impairment, even though the probability is low. (53)

Neonatologists counseling caregivers of neonates at risk for CP should be aware of the potential for future direct negative ramifications on the caregivers. Children who have long-term functional limitations, such as with CP, require a high level of care, which may have a negative impact on the physical and mental health of the caregivers compared with the health of caregivers of children who are not so affected. (61)(62) Caregivers of children who have CP have higher levels and chronicity of distress, emotional and cognitive problems, physical problems, and financial strain compared with the general population. (61) When their child is diagnosed with CP, mothers may experience loss of both an “ideal” child and expectations of “normal” motherhood. (63) Early discussions in the NICU and in high-risk infant follow-up clinics informing caregivers of the possible negative impacts of caring for a child who has CP may allow for early lifestyle adjustments through potential management strategies to minimize stress.

Counseling caregivers on future adverse developmental risks can be challenging, especially because the cause of CP remains unexplained in most cases. More serious counseling should be given to the parents of infants who are the most likely to develop CP based on abnormal neuroimaging findings and other risk factors associated with CP (Table 2).

Himpens and colleagues (64) recently proposed a predictive model based on perinatal characteristics and neonatal ultrasonographic-detected brain injuries to identify specific infants at risk for developing CP. There is no single perfect approach to conveying serious, life-changing, and typically overwhelming medical information to caregivers, but certain strategies striving for improved communication are worthy of consideration. Ideally, counseling should involve both parents when possible or someone who can provide support to a single parent and in a setting with minimal interruptions that allows for privacy immediately after the counseling. (58)(65) Neonatologists should be direct, clear, sympathetic, honest, and open, with a balanced viewpoint when discussing the potential for future impairments; acknowledge the limitations of accurate predictions given current diagnostic capabilities; and allow ample time for parents to ask questions. (58)(65) Providing sufficient information has been very important when assessing levels of parental satisfaction with disclosure of a disability. (66) Parents should be offered the chance for timely further discussion of potential future diagnoses with the professionals present at the original interview. Providing parents with resources (written information or trusted web sites) on CP and early support programs may improve their understanding and satisfaction with their child’s health care. The availability of professional support from a social worker, psychologist, or other health-care worker at the time of counseling, particularly when a formal diagnosis of a life-impacting disability such as CP is made, may help accommodate parental needs in adjusting to unfavorable news. Keeping parents of infants at high risk for the development of long-term disabilities well-informed in the NICU, before the formal diagnosis of CP, may lead to better long-term parental coping mechanisms and subsequent better care for the affected child.

Table 2. Characteristic Risk Factors Associated With Cerebral Palsy Prevalence Rates

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<tr>
<th>Increased Prevalence</th>
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<tr>
<td>● Deprived socioeconomic populations</td>
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<td>● Male sex</td>
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<td>● Racial disparity: In the United States, prevalence highest among African American males</td>
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<td>● Preterm premature rupture of membranes</td>
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<td>● Low Apgar scores</td>
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<td>● Perinatal asphyxia</td>
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<td>● Mechanical ventilation &gt; 7 d</td>
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<td>Decreased Prevalence</td>
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<tr>
<td>● Preeclampsia</td>
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<td>● Antenatal magnesium sulphate therapy</td>
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<td>● Antenatal corticosteroids</td>
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<td>● Caffeine: neonatal therapy</td>
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- Know the indications for and limitations of various neuroimaging studies and be able to recognize normal and abnormal structures and changes during development and growth.
- Know the approximate risk of cerebral palsy in very low birth weight, moderately low birth weight, and normal birth weight infants.
- Know the prenatal, perinatal, and neonatal risk factors for the development of cerebral palsy.

References

7. Rosenbaum P. The definition and classification of cerebral palsy: are we any further ahead in 2006? *NeuroReviews*. 2006;17:e569–e574
1. The current definition of cerebral palsy (CP) was published in 2005 based on the conclusions of an international workshop of experts in the field of developmental disorders. Of the following, the most accurate statement regarding CP is that it:

A. Involves aberrations of movement and posture.
B. Is a progressive neurodevelopmental disorder.
C. Is viewed as a homogeneous disorder of a single cause.
D. Originates during early childhood after infancy.
E. Spares disturbances of sensation and behavior.

2. A 14-week-old infant, who weighed 610 g at birth at an estimated gestational age of 24 weeks, is being discharged from the neonatal intensive care unit. Scheduling for neurodevelopmental follow-up of the infant is being arranged. The mother is concerned about possible CP and inquires about when such a diagnosis is usually made. Of the following, the definitive diagnosis of cerebral palsy is typically made after the corrected age of:

A. 6 months.
B. 12 months.
C. 18 months.
D. 24 months.
E. 36 months.

3. According to a population-based follow-up study using the Medical Birth Registry of Norway, the prevalence of CP among infants born at term is estimated at 1.15 per 1,000 births. The risk of CP in these infants varies, depending on the timing of birth within the term period. Of the following, the gestational age at birth within the term period most associated with a reduced risk of CP is:

A. 37 weeks.
B. 38 weeks.
C. 39 weeks.
D. 40 weeks.
E. 41 weeks.

4. Abnormal neuroanatomic findings are seen on neuroimaging in 80% to 90% of children who have CP. Of the following, the most common neuroanatomic abnormality among such children involves the:

A. Basal ganglia.
B. Commissural fibers.
C. Limb of internal capsule.
D. Periventricular white matter.
E. Superior corona radiata.
5. Cerebral palsy is categorized into phenotypic subtypes based on the distribution of limb involvement and type of muscle tone abnormality. Of the following, the most common phenotype of CP among preterm infants is:

A. Ataxic hemiplegia.
B. Athetoid monoplegia.
C. Dyskinetic quadriparesis.
D. Epileptiform dyskinesia.
E. Spastic diplegia.

6. Early counseling of the parents of a neonate at high risk for CP may function as the starting point for parental adaptation to their infant’s lifelong condition and its associated morbidities. Of the following, the most frequent morbidity associated with CP is:

A. Dental caries.
B. Epilepsy.
C. Feeding problems.
D. Speech impairment.
E. Stunted growth.
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Electronic Fetal Monitoring Case Review Series

Electronic fetal monitoring (EFM) is a popular technology used to establish fetal well-being. Despite its widespread use, terminology used to describe patterns seen on the monitor has not been consistent until recently. In 1997, the National Institute of Child Health and Human Development (NICHD) Research Planning Workshop published guidelines for interpretation of fetal tracings. This publication was the culmination of 2 years of work by a panel of experts in the field of fetal monitoring and was endorsed in 2005 by both the American College of Obstetricians and Gynecologists (ACOG) and the Association of Women’s Health, Obstetric and Neonatal Nurses (AWHONN). In 2008, ACOG, NICHD, and the Society for Maternal-Fetal Medicine reviewed and updated the definitions for fetal heart rate patterns, interpretation, and research recommendations. Following is a summary of the terminology definitions and assumptions found in the 2008 NICHD workshop report. Normal values for arterial umbilical cord gas values and indications of acidosis are defined in Table 1.

Assumptions from the NICHD Workshop

- Definitions are developed for visual interpretation, assuming that both the fetal heart rate (FHR) and uterine activity recordings are of adequate quality
- Definitions apply to tracings generated by internal or external monitoring devices
- Periodic patterns are differentiated based on waveform, abrupt or gradual (e.g., late decelerations have a gradual onset and variable decelerations have an abrupt onset)
- Long- and short-term variability are evaluated visually as a unit
- Gestational age of the fetus is considered when evaluating patterns
- Components of fetal heart rate FHR do not occur alone and generally evolve over time

Definitions

Baseline Fetal Heart Rate

- Approximate mean FHR rounded to increments of 5 beats/min in a 10-minute segment of tracing, excluding accelerations and decelerations, periods of marked variability, and segments of baseline that differ by >25 beats/min
- In the 10-minute segment, the minimum baseline duration must be at least 2 minutes (not necessarily contiguous) or the baseline for that segment is indeterminate
- Bradycardia is a baseline of <110 beats/min; tachycardia is a baseline of >160 beats/min
- Sinusoidal baseline has a smooth sine wave-like undulating pattern, with waves having regular frequency and amplitude

Baseline Variability

- Fluctuations in the baseline FHR of two cycles per minute or greater, fluctuations are irregular in amplitude and frequency, fluctuations are visually quantitated as the amplitude of the peak to trough in beats per minute
- Classification of variability:
  - Absent: Amplitude range is undetectable
  - Minimal: Amplitude range is greater than undetectable to 5 beats/min
  - Moderate: Amplitude range is 6 to 25 beats/min
  - Marked: Amplitude range is >25 beats/min
Accelerations

- Abrupt increase in FHR above the most recently determined baseline
- Onset to peak of acceleration is <30 seconds, acme is ≥15 beats/min above the most recently determined baseline and lasts ≥15 seconds but <2 minutes
- Before 32 weeks’ gestation, accelerations are defined by an acme ≥10 beats/min above the most recently determined baseline for ≥10 seconds
- Prolonged acceleration lasts ≥2 minutes but <10 minutes

Late Decelerations

- Gradual decrease in FHR (onset to nadir ≥30 seconds) below the most recently determined baseline, with nadir occurring after the peak of uterine contractions
- Considered a periodic pattern because it occurs with uterine contractions

Early Decelerations

- Gradual decrease in FHR (onset to nadir ≥30 seconds) below the most recently determined baseline, with nadir occurring coincident with uterine contraction
- Also considered a periodic pattern

Variable Decelerations

- Abrupt decrease in FHR (onset to nadir <30 seconds)
- Decrease is ≥15 beats/min below the most recently determined baseline lasting ≥15 seconds but <2 minutes
- May be episodic (occurs without a contraction) or periodic

Prolonged Decelerations

- Decrease in the FHR ≥15 beats/min below the most recently determined baseline lasting ≥2 minutes but <10 minutes from onset to return to baseline

Decelerations are tentatively called recurrent if they occur with ≥50% of uterine contractions in a 20-minute period.

Decelerations occurring with <50% of uterine contractions in a 20-minute segment are intermittent.

Sinusoidal Fetal Heart Rate Pattern

- Visually apparent, smooth sine wavelike undulating pattern in the baseline with a cycle frequency of 3 to 5/minute that persists for ≥20 minutes.

Uterine Contractions

- Quantified as the number of contractions in a 10-minute window, averaged over 30 minutes.
  - Normal: ≤5 contractions in 10 minutes
  - Tachysystole: >5 contractions in 10 minutes

Interpretation

A three-tier Fetal Heart Rate Interpretation system has been recommended as follows:

- Category I FHR tracings: Normal, strongly predictive of normal fetal acid-base status and require routine care. These tracings include all of the following:
  - Baseline rate: 110 to 160 beats/min
  - Baseline FHR variability: Moderate
  - Late or variable decelerations: Absent
  - Early decelerations: Present or absent
  - Accelerations: Present or absent

- Category II FHR tracings: Indeterminate, require evaluation and continued surveillance and reevaluation. Examples of these tracings include any of the following:
  - Bradycardia not accompanied by absent variability
  - Tachycardia
  - Minimal or marked baseline variability
  - Absent variability without recurrent decelerations
  - Absence of induced accelerations after fetal stimulation
  - Recurrent variable decelerations with minimal or

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moderate variability

- Prolonged decelerations
- Recurrent late decelerations with moderate variability
- Variable decelerations with other characteristics, such as slow return to baseline

- Category III FHR tracings: Abnormal, predictive of abnormal fetal acid-base status and require prompt intervention. These tracings include:
  - Absent variability with any of the following:
    - Recurrent late decelerations
    - Recurrent variable decelerations
    - Bradycardia
  - Sinusoidal pattern


We encourage readers to examine each strip in the case presentation and make a personal interpretation of the findings before advancing to the expert interpretation provided.
Case Presentation

History

A 38-year-old G2P1001 at 37 6/7 weeks gestation presents to labor and delivery from clinic after she had two elevated blood pressures greater than 140/90 and trace proteinuria. She also has a headache and new onset upper extremity swelling. She denies any visual changes or right upper quadrant pain. She has irregular uterine contractions and active fetal movement. She denies leaking of fluid or vaginal bleeding. Her pregnancy is complicated by advanced maternal age. She declined genetic screening. Her prenatal laboratory tests are all normal.

Case Progression

On physical examination, her blood pressure is 140/103 mm Hg and she has a urine dipstick of 1+(30 mg/dL) of protein. She meets the criteria for severe preeclampsia based on her headache. (1) Her cervix is 30% effaced, 1 cm dilated, and negative 3 station. All laboratory values were normal, except for a slightly increased AST of 45 U/L. A decision is made to proceed with induction of labor secondary to severe preeclampsia. A fetal heart tracing is obtained on admission (Fig. 1).

Figure 1. EFM strip #1.
Findings from electronic fetal monitoring (EFM) strip #1 are:

- Variability: Moderate
- Baseline rate: 135 beats/min
- Episodic Pattern: Acceleration
- Periodic Pattern: None
- Uterine Contractions: Every 5 minutes
- Interpretation: Category I tracing
- Differential Diagnosis: Normal fetal tracing.

- Action: No intervention required. The patient is admitted for induction of labor.

A Foley balloon was placed for cervical ripening. Four hours later, the balloon is expelled and she is found to be 50%/5 cm/−2. She is started on pitocin, and 1 hour later she received an epidural for pain control. She is examined 8 hours later and is found to be 60%/6 cm/−1. She undergoes artificial rupture of membranes and has light meconium. The fetal tracing is shown in Figure 2.
Findings from EFM Strip #2 are:

- Variability: Moderate
- Baseline rate: 120 beats/min
- Episodic Pattern: None
- Periodic Pattern: None observed
- Uterine Contractions: Every 3 minutes
- Interpretation: Category I Tracing
- Differential Diagnosis: Normal fetal tracing. The maternal pulse oximeter demonstrates that the maternal heart rate also occasionally is 120 beats/min. It is important to distinguish the maternal and fetal heart rates.
- Action: Overall, the tracing is reassuring due to moderate variability. Induction of labor can continue.

The pitocin is continued and 4 hours later she remains 6 cm dilated. An intrauterine pressure catheter is placed to assess the uterine contractions. The fetal tracing is shown in Figure 3.
Findings from EFM Strip #3 are:

- Variability: Moderate
- Baseline Rate: 120 beats/min
- Episodic Pattern: Accelerations
- Periodic Pattern: None
- Uterine Contractions: Occurring every 3 minutes, lasting 50 seconds
- Interpretation: Category I tracing
- Differential Diagnosis: Normal tracing, with evidence of fetal well-being indicated by moderate variability and accelerations. The patient may not be progressing due to inadequate uterine contractions. With the placement of the intrauterine pressure catheter, the uterine power can be documented.

Action: No intervention required.

Three hours later, the patient is re-evaluated. No cervical exam is performed. The fetal heart tracing is shown below (Fig. 4).
Findings from EFM Strip #4 are:

- Variability: Moderate
- Baseline rate: 125 beats/min
- Episodic Pattern: Accelerations
- Periodic Pattern: None
- Uterine Contractions: Occurring every 2 minutes
- Interpretation: Category I tracing

- Differential Diagnosis: Normal tracing, with evidence of fetal well-being indicated by moderate variability and accelerations.
- Action: No intervention required.

Ninety minutes later, the patient has increasing pain with contractions. Her cervical examination is 90%/9/0. Another fetal heart tracing is shown below (Fig. 5).
Findings from EFM Strip #5 are:

- Variability: Minimal
- Baseline rate: 150 beats/min
- Episodic Pattern: None
- Periodic Pattern: Recurrent variable decelerations
- Uterine Contractions: Occurring every 3–4 minutes
- Interpretation: Category II
- Differential Diagnosis: The differential diagnosis includes umbilical cord compression, uteroplacental underperfusion or uteroplacental dysfunction. Variable decelerations are normally well tolerated, however if they continue to be repetitive or more severe, acidemia can develop.

- Action: The fetal status should be monitored closely.

Ninety minutes later, the physician is called to the room for evaluation of the fetal heart rate tracing. Her cervix is unchanged. The fetal tracing is shown below (Fig. 6).
Findings from EFM Strip #6 are:

- Variability: Minimal/moderate
- Baseline Rate: 140 rising to 150 beats/min
- Episodic Pattern: None
- Periodic Pattern: Variable deceleration and possible sinusoidal pattern
- Uterine Contractions: None documented
- Interpretation: Category III
- Differential Diagnosis: Sinusoidal patterns can be associated with fetal anemia, use of certain drugs, fetal asphyxia/hypoxia, fetal infection, fetal cardiac anomalies, and fetal sleep cycles. This patient had not received any new medications.

- Action: Due to the concerning fetal heart tracing, immediate delivery is indicated.

Decision is made to proceed with cesarean delivery due to the sinusoidal heart rate pattern. The fetal heart rate tracing in the operating room is shown below (Fig. 7).

Figure 6. EFM strip #6.

Figure 7. EFM strip #7.
Findings from EFM Strip #7 are:

- Variability: Absent
- Baseline Rate: 160 beats/min
- Episodic Pattern: Sinusoidal heart rate pattern
- Periodic Pattern: None
- Uterine Contractions: None documented
- Interpretation: Category III
- Differential Diagnosis: Unchanged
- Action: Unchanged

The patient’s epidural was not adequate after a bolus was infused; therefore, she received general anesthesia. Twenty-eight minutes from the decision for cesarean, the baby is delivered. There is an abruption noted upon entry into the uterine cavity. The patient is admitted to the intensive care unit secondary to the general anesthesia and abruption. Her international normalized ratio (INR) is elevated at 1.36; however, she recovers and is transferred to the mother–baby unit on postoperative day 1.

Outcome

A viable male infant is delivered by cesarean section. He weighs 2,880 g (6 lb; 5.6 oz) and has Apgar scores of 1 at 1 minute, 3 at 5 minutes, 5 at 10 minutes, and 7 at 15 minutes. The blood gas is shown below. This is consistent with a mixed acidosis. A large blood clot is cleared from his throat, and bloody mucus is suctioned from his throat following intubation. He receives positive pressure ventilation (PPV) and chest compressions and responds appropriately. He is placed on continuous positive airway pressure for 30 minutes and then is weaned to room air. His hemoglobin is 19.1 g/dL, which is normal. He is observed in the neonatal intensive care unit for 48 hours and then transferred to the patient’s room on day 2 after birth and is discharged home on day 4.

Placental abruption is the premature separation of the placenta before delivery, which occurs in approximately 1 in 100 deliveries. It is a known complication of severe preeclampsia. (2) It is a clinical diagnosis usually consisting of vaginal bleeding, abdominal pain, increased uter-

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ine tone, tachysystole, and a nonreassuring fetal heart rate tracing. In 10% to 20% of abruptions, there may be no vaginal bleeding. (3) The nonreassuring fetal heart rate tracing can consist of late decelerations, minimal variability, fetal tachycardia, or, as in our patient, a sinusoidal pattern. A sinusoidal heart rate pattern by definition is a visually apparent, smooth sine wavelike undulating pattern in the baseline with a cycle frequency of 3 to 5 minutes that persists for ≥20 minutes. This pattern has been seen with fetal anemia, use of certain drugs, fetal asphyxia/hypoxia, fetal infection, fetal cardiac anomalies, and fetal sleep cycles. (4) A true sinusoidal pattern can be an ominous sign of fetal jeopardy and prompt intervention is required.

References

Correction
In the article entitled “Cerebral Palsy: Prevalence, Predictability, and Parental Counseling” in the October issue (NeoReviews 2011;12:e564–e574), at the bottom of page e565, the text incorrectly states: “The bilateral spastic form increased from 0.58 (95% confidence interval [CI], 0.41 to 0.80) in 1980 to 0.33 (CI, 0.22 to 0.46) in 1998 . . .” This sentence should read “The bilateral spastic form decreased from 0.58 to 0.33 . . .”. The journal regrets the error.