An Algorithm for Identifying and Classifying Cerebral Palsy in Young Children

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Objective To develop an algorithm on the basis of data obtained with a reliable, standardized neurological examination and report the prevalence of cerebral palsy (CP) subtypes (diparesis, hemiparesis, and quadriparesis) in a cohort of 2-year-old children born before 28 weeks gestation.

Study design We compared children with CP subtypes on extent of handicap and frequency of microcephaly, cognitive impairment, and screening positive for autism.

Results Of the 1056 children examined, 11.4% (120) were given an algorithm-based classification of CP. Of these children, 31% had diparesis, 17% had hemiparesis, and 52% had quadriparesis. Children with quadriparesis were 9 times more likely than children with diparesis (76% versus 8%) to be more highly impaired and 5 times more likely than children with diparesis to be microcephalic (43% versus 8%). They were more than twice as likely as children with diparesis to have a score <70 on the mental scale of the BSID-II (75% versus 34%) and had the highest rate of the Modified Checklist for Autism in Toddlers positivity (76%) compared with children with diparesis (30%) and children without CP (18%).

Conclusion We developed an algorithm that classifies CP subtypes, which should permit comparison among studies. Extent of gross motor dysfunction and rates of co-morbidities are highest in children with quadriparesis and lowest in children with diparesis. *(J Pediatr 2008;153:466-72)*

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Cerebral palsy (CP) is a group of non-progressive permanent disorders of movement and posture that occur following damage to the developing fetal or infant brain. It is often accompanied by other neurodevelopmental disorders.[1-3] CP occurs in 0.2% of live births, but infants born before 28 weeks gestation have a 50-fold elevated risk when compared with infants born at term,[4] with a prevalence between 6% and 26%.[5-12]

Part of the variability in prevalence may be attributable to the lack of a published operational identification or classification of CP that can be used and replicated by clinicians across settings. Because of inconsistencies in identifying forms of CP, some experts have recommended classifying CP primarily on the basis of the degree of severity of gross motor function, while minimizing or eliminating classic topography-based categorization of CP types.[13-15] In response, we created an algorithm to more reliably identify CP and topography-based subtypes of CP that could be replicated by others. The decision tree of the algorithm we developed models the way a seasoned pediatric neurology clinician might identify and classify CP.

We sought additional confirmation that the CP subtypes identified by our algorithm were distinctive with respect to the clinical severity of dysfunction and in the frequency of associated abnormal findings. We anticipated that children with quadriparesis would be most highly affected or have greater numbers of co-morbid conditions, followed by children with diparesis, children with hemiparesis, and children with no CP. Specifically, we sought to determine the extent to which children with different subtypes varied in: 1) their levels of dysfunction as assessed by the Gross Motor Functional

CP	Cerebral palsy	M-CHAT	Modified Checklist for Autism in Toddlers
ELGAN	Extremely Low Gestational Age Newborns	MDI	Mental scale of the BSID-II
GMFCS	Gross Motor Functional Classification Scale	PDI	Motor scales of the BSID-II

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Classification Scale (GMFCS) and 2) their frequency of microcephaly, cognitive impairment, and positive screening on the Modified Checklist for Autism in Toddlers (M-CHAT) at 2 years adjusted age.

METHODS

ELGAN Study

The Extremely Low Gestational Age Newborns (ELGAN) study was designed to identify characteristics and exposures that increase the risk of disorders of brain structure and function (including CP) in extremely low gestational age newborns. From 2002 to 2004, women delivering before 28 weeks gestation at 1 of 14 participating institutions in 11 cities in 5 states were asked to enroll in the study. Of the 1201 surviving newborns, 1056 (88%) underwent neurological examinations at 2 years of age and are the subject of this report. This study was approved by all involved institutional human studies review boards, and all families consented to participate in the study.

To standardize neurological examinations across all sites, a stand-alone, multimedia-training video/CD-ROM was developed,[16] on the basis of elements of a standard neurological examination.[17-20] Use of the video-CD led to a reliability of 88% to 96% when examiner findings were compared with a gold standard assessment.[16] Examiners also evaluated level of disability by using the GMFCS.[14,21-23] Neurological examiners remained largely unaware of the child's specific medical history, other than that the infant had extremely low gestational age at birth.

Certified examiners administered and scored the mental (MDI) and motor (PDI) scales of the BSID-II. Before testing, examiners were told only the child's age. After test completion, they were told the gestational age to adjust the MDI and PDI for the degree of prematurity. Of the 1056 children who were identified to have CP on the basis of the algorithm, 59 were considered not testable for the MDI and 76 for the PDI. We used the Vineland Adaptive Behavioral Composite scale as a proxy for the MDI for 39 children and the Vineland Motor Skills domain scale as a proxy for the PDI for 43 children. Caregivers of study participants completed the Modified Checklist for Autism in Toddlers (M-CHAT) screen survey.[24]

Neurological Examination Instrument

The data collection form included 7 items in the upper extremities in 4 areas of motor function: motor strength (4 items), tone alteration (1 item), posture (1 item), and hand use (1 item). Two areas of function were evaluated in the lower extremities: strength (2 items) and tone (3 items). In our strength assessment, we use indirect measures of power, including the child's ability to push the chest up off the bed with the arms, support body weight on the legs, and lift and move arms and legs.

Algorithm Assumptions

Assumption 1. An algorithm that simplifies options is most useful. The range of presentations of topography-based classification of diparesis, quadriparesis, and hemiparesis can include partial forms. For example, monoparesis also occurs. Rather than create a category for monoparesis, we viewed monoparesis of an upper extremity as a partial hemiparesis. Our final categorization includes these 3 groups:

- Quadriparesis: involvement of both lower extremities and involvement of 1 (asymmetric) or both (symmetric) upper extremities; or involvement of both upper extremities and 1 lower extremity (asymmetric quadriparesis);
- Diparesis: involvement of both lower extremities only or only 1 leg;
- Hemiparesis: involvement restricted to only 1 side of the body.

Assumption 2. Dystonia and dyskinetic forms of CP are more evident later. We did not distinguish qualitative forms of abnormally elevated tone (hypertonia), particularly spasticity and dystonia. Dystonia and spasticity co-occur frequently, and the presence of spasticity may make identification of dystonia more difficult.[25,26] The distinction between the 2 also may be difficult because signs of dystonia may be intermittent and vary with state and level of activity. Finally, the expression of dystonia and dyskinetic forms of CP evolves in the first years of life and usually manifests more obviously later.[27]

Assumption 3. The proposed algorithm's value may be limited to the very young child born extremely premature performed at 2 years corrected age. The examination we used and the proposed algorithm was tested and applied to children in the first few years of life. CP evolves in its presentation, sometimes becoming more complex in later years. For example, choreoathetosis, more often seen in infants born at term, becomes more obvious after the first years of life. Because motor findings characteristic of CP can improve or dissipate at later ages,[28-30] we can expect some children given a CP diagnosis at a young age, whether algorithm-based or not, to no longer be given the same diagnosis years later.

Algorithm development as an iterative process: In analyzing the CD-based neurological examination findings, a number of decisions were made sequentially (Figure).

First, components of the examination that did not specifically evaluate motor status were excluded (eg, visual interactions, extra-ocular muscles).

Second, because the evaluation of deep tendon reflexes is less reliably assessed than other parts of the examination and probably less specific to motor impairment, an effort was made to minimize their impact on the decision tree. After considering approaches that assigned less weight to deep tendon reflexes, we decided to exclude this item from the decision tree.

Third, we required multiple, corroborating abnormal findings. Although we preferred the presence of at least 2 abnormal findings that assess different domains of the motor