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Practice Parameter: Diagnostic assessment of the child with cerebral palsy

Report of the Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurology Society

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Abstract—Objective: The Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurology Society develop practice parameters as strategies for patient management based on analysis of evidence. For this parameter the authors reviewed available evidence on the assessment of a child suspected of having cerebral palsy (CP), a nonprogressive disorder of posture or movement due to a lesion of the developing brain. **Methods:** Relevant literature was reviewed, abstracted, and classified. Recommendations were based on a four-tiered scheme of evidence classification. **Results:** CP is a common problem, occurring in about 2 to 2.5 per 1,000 live births. In order to establish that a brain abnormality exists in children with CP that may, in turn, suggest an etiology and prognosis, neuroimaging is recommended with MRI preferred to CT (Level A). Metabolic and genetic studies should not be routinely obtained in the evaluation of the child with CP (Level B). If the clinical history or findings on neuroimaging do not determine a specific structural abnormality or if there are additional and atypical features in the history or clinical examination, metabolic and genetic testing should be considered (Level C). Detection of a brain malformation in a child with CP warrants consideration of an underlying genetic or metabolic etiology. Because the incidence of cerebral infarction is high in children with hemiplegic CP, diagnostic testing for coagulation disorders should be considered (Level B). However, there is insufficient evidence at present to be precise as to what studies should be ordered. An EEG is not recommended unless there are features suggestive of epilepsy or a specific epileptic syndrome (Level A). Because children with CP may have associated deficits of mental retardation, ophthalmologic and hearing impairments, speech and language disorders, and oral-motor dysfunction, screening for these conditions should be part of the initial assessment (Level A). **Conclusions:** Neuroimaging results in children with CP are commonly abnormal and may help determine the etiology. Screening for associated conditions is warranted as part of the initial evaluation.

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Cerebral palsy (CP) can be defined as a disorder of aberrant control of movement and posture, appearing early in life secondary to a CNS lesion or dysfunction that is not the result of a recognized progressive or degenerative brain disease.¹ The brain abnormality

may occur pre-, peri-, or postnatally. The diagnosis of CP always involves a motor deficit and the usual presenting complaint for which medical evaluation is sought is that the child is not reaching motor milestones at the appropriate chronological age. In most

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instances, a medical history establishes that the child is not losing function, assuring that the patient does not have a progressive disease. This history, combined with a neurologic examination establishing that the patient's motor deficit is due to a cerebral abnormality, leads to the diagnosis of CP. Serial examinations may be necessary to assure the diagnosis of CP, especially when the history is not reliable.

There is agreement that CP is due to a defect or lesion in the developing brain, which may have had its onset in the prenatal, perinatal, or postnatal period.¹ While often a cutoff age for the appearance of symptoms early in life is generally not given, the great majority of children with CP present with symptoms as infants or toddlers, and the diagnosis of CP is made before age 2 years. In some children, symptom onset may be delayed (e.g., dystonic cerebral palsy), whereas in other children the appearance of pseudoprogession of symptoms may occur. The term CP is descriptive and includes a number of etiologies and clinical presentations. Although there is no consensus about a precise age cut-off either for the timing of the insult or the onset of symptoms, of importance is that affected individuals have similar needs for rehabilitation, education, and medical and social services.

CP is a common problem. The worldwide incidence of CP is approximately 2 to 2.5 per 1,000 live births.² Each year about 10,000 babies born in the United States develop CP.³ CP occurs more commonly in children who are born very prematurely or at term. Data from Sweden on 241 children with CP indicate that 36% were born at a gestational age (GA) of less than 28 weeks; 25% at 28 to 32 weeks GA; 2.5% at 32 to 38 weeks GA; and 37% at term.²

The types and severity of CP are clinically well established. A Scandinavian study reported that 33% of the CP population was hemiplegic, 44% diplegic, and 6% quadriplegic.²

Half of the individuals with CP use assistive devices such as braces, walkers, or wheelchairs to help develop or maintain mobility and almost 70% have other disabilities, primarily mental retardation.³ Individuals with CP may need specialized medical care, educational and social services, and other help throughout their lives from their families and communities. A study 10 years ago in California of the extra economic costs associated with CP and 17 other congenital disorders (e.g., Down syndrome, spina bifida) showed that CP had the highest lifetime costs per new case, averaging \$503,000 in 1992 dollars.⁴

Laboratory tests are not necessary to confirm the diagnosis of CP as it is based on the history and examination. It is a syndrome with many etiologies. Genetic diseases, brain malformations, infections, and anoxic injury to the developing brain are a few of the many causes of CP. Accurate determination of the etiology of CP has specific implications regarding treatment, prognosis, and ongoing medical management of associated conditions. The importance of de-

termining whether there is a malformation, genetic etiology, or injury and whether the injury is due to an acquired pre-, peri-, or postnatal process has obvious significance from the point of view of assessment of recurrence risk, counseling of families, and implementation of prevention programs, and when medicolegal issues arise. Determining causality also helps limit further unnecessary testing. Finally, understanding the etiology of CP has implications for prevention and intervention strategies. This knowledge will direct further research efforts.

Determining the types of evaluations appropriate for children with CP has posed tremendous challenges for parents and health care providers. The purpose of this practice parameter is to review data regarding the value and role of diagnostic tests used to evaluate children diagnosed with CP. Data regarding the role of neuroimaging, metabolic and genetic testing, and evaluation for coagulopathy are discussed. There was insufficient evidence to make any recommendations regarding the role of SPECT scans or evoked potentials in children with CP. This parameter also reviews evidence regarding the prevalence of associated problems such as epilepsy, mental retardation, speech and language disorders, and ophthalmologic and hearing impairments, and the need for their systematic evaluation.

Description of process. Literature searches were conducted with the assistance of the University of Minnesota Biomedical Information Services for relevant articles published from 1980 to March 2002. Medline, CINAHL, and Healthstar databases were searched for relevant articles published from 1966 to 2002, in the English language, using the following key words: CP, magnetic resonance imaging, MRI, computed axial tomography, CT scan, single photon emission tomography, SPECT, metabolic disease, thrombophilia, brain stem evoked potentials, sensory evoked potentials, visual evoked potentials, electroencephalography (EEG), seizures, epilepsy, vision loss, hearing loss, developmental delay, and speech and language delay. Approximately 350 titles and abstracts were reviewed for content regarding the establishment of the etiology of CP. Articles were excluded if the tests were ordered for reasons other than to establish the etiology. We included only studies that contained more than 20 patients; smaller case series were excluded. The ages of infants and children included in these studies were similar to the ages of children typically seen for diagnostic evaluation so it was believed that the evidence-based recommendations included in this parameter were appropriate. It was also believed that as CP is usually due to a static process, it was unlikely for neuroimaging studies to change over time so that data from studies done in older children with CP were valid regarding etiologic yield. Each article was reviewed, abstracted, and classified by a committee member. Data extracted include first author, year, study population, study design, number of patients, types of CP, results of testing,

Table 1 American Academy of Neurology evidence classification scheme for determining the yield of established diagnostic and screening tests

Class I: A statistical,¹ population-based² sample of patients studied at a uniform point in time (usually early) during the course of the condition. All patients undergo the intervention of interest. The outcome, if not objective,⁵ is determined in an evaluation that is masked to the patients' clinical presentations.

Class II: A statistical, non-referral-clinic-based³ sample of patients studied at a uniform point in time (usually early) during the course of the condition. Most (>80%) patients undergo the intervention of interest. The outcome, if not objective,⁵ is determined in an evaluation that is masked to the patients' clinical presentations.

Class III: A selected, referral-clinic-based⁴ sample of patients studied during the course of the condition. Some patients undergo the intervention of interest. The outcome, if not objective,⁵ is determined in an evaluation by someone other than the treating physician.

Class IV: Expert opinion, case reports, or any study not meeting criteria for class I to III.

This is a new classification scheme developed by the Quality Standards Subcommittee (QSS) for studies related to determining the yield of established diagnostic and screening tests or interventions and is appropriate only when the diagnostic accuracy of the test or intervention is known to be good. Additionally, the abnormality potentially identified by the screening intervention should be treatable or should have important prognostic implications. This classification is different from others currently recommended by the QSS that have been published in recent parameters that relate to diagnostic, prognostic, or therapeutic studies.

¹ Statistical sample: A complete (consecutive), random, or systematic (e.g., every third patient) sample of the available population with the disease;

² Population-based: The available population for the study consists of all patients within a defined geographic region;

³ Non-referral-clinic-based: The available population for the study consists of all patients presenting to a primary care setting with the condition;

⁴ Referral-clinic-based: The available population for the study consists of all patients referred to a tertiary care or specialty setting. These patients may have been selected for more severe or unusual forms of the condition and thus may be less representative;

⁵ Objective: An outcome measure that is very unlikely to be affected by an observer's expectations (e.g., determination of death, the presence of a mass on head CT, serum B12 assays).

and outcomes measured. A new four-tiered classification scheme for determining the yield of established diagnostic and screening tests developed by the Quality Standards Subcommittee was utilized as part of this assessment (table 1). This classification scheme is different from the one currently used in recently published practice parameters that evaluate diagnostic, prognostic, or therapeutic articles. Depending on the strength of this evidence, it was decided whether specific recommendations could be made, and if so, the level of strength of these recommendations (table 2).

Neuroimaging. Should neuroimaging be routinely obtained in the child with CP?

Table 2 American Academy of Neurology system for translation of evidence to recommendations

Rating of recommendations	Translation of evidence to recommendations
A = Established as useful/predictive or not useful/predictive for the given condition in the specified population	Level A rating requires at least one convincing class I study or at least two consistent, convincing class II studies
B = Probably useful/predictive or not useful/predictive for the given condition in the specified population	Level B rating requires at least one convincing class II study or overwhelming class III evidence
C = Possibly useful/predictive or not useful/predictive for the given condition in the specified population	Level C rating requires at least two convincing class III studies
U = Data inadequate or conflicting. Given current knowledge, test, predictor is unproven.	

Evidence. In neonates, neuroimaging is frequently obtained when there is a history of complications during pregnancy, labor, and delivery; when the infant is born very prematurely (<32 weeks); or when neurologic symptoms or findings are present on neonatal examination. A practice parameter regarding the value of and indications for obtaining neuroimaging in preterm and term infants has recently been published.⁵ The current parameter addresses the role of neuroimaging in the infant or child who has been diagnosed with or is suspected of having CP based upon a motor deficit or delay that is not worsening and a history and neurologic examination that places the lesion in the CNS. In some situations, the risks of obtaining a neuroimaging study in an infant with CP may potentially outweigh the benefits of further defining the etiology.

Recent data encompassing a total of 1,426 children who underwent either CT⁶⁻¹⁴ or MRI¹⁵⁻²⁴ scans indicated an abnormality in 62% to 100% of individuals (mean for CT, 77%; for MRI, 89%) (tables 3a and 4a). For the combined CT and MRI class I studies (n = 238), 88% of children had abnormal scans whereas 77% of patients in a class II study (n = 22) had abnormal scans as did 83% of children in the class III studies (n = 1166). The timing of the brain insult causing CP may be categorized into prenatal, perinatal, and postnatal subgroups and the neuropathologic changes seen in CP have been linked to the gestational age of the infant at the time of insult. Historical features (e.g., neonatal intracranial hemorrhage, neonatal encephalopathy, stroke, CNS infection, trauma) or prior neuroimaging studies that document such disorders or the presence of a brain malformation may provide sufficient information to establish the etiology when a child is seen later in life as the symptoms of CP evolve. Likewise, findings

Table 3 CT in children with cerebral palsy (CP)

a. Overall yield of finding an abnormal CT scan in children with CP

Reference	Class	N	Ages, y	Type of CP	% Abnormal
Wiklund et al., 1991 ⁶	I	83	5–16	H	73
Wiklund et al., 1991 ⁷	I	28	5–16	H	75
Miller and Cala, 1989 ⁸	I	29	6–35	A	62
Chen, 1981 ⁹	III	281	0.08–7	M	84
Kolawole et al., 1989 ¹⁰	III	120	1–>10	M	73
Taudorf et al., 1984 ¹¹	III	83	NA	M	67
Schouman-Claeys et al., 1989 ¹²	III	76	0.6–15	M	63
Cohen and Duffner, 1981 ¹³	III	52	0.67–>10	H	87
Molteni et al., 1987 ¹⁴	III	30	5–16	H	93
Total no. of patients		782			
Mean abnormal CT					77

b. Percent of patients with an abnormal CT scan based on the type of CP (n = 485)

Hemiplegic (n = 146)	89	Quadriplegic (n = 111)	70
Ataxic (n = 19)	88	Hypotonic (n = 19)	73
Mixed (n = 29)	79	Dyskinetic (n = 14)	36
Diplegic (n = 153)	75		

c. Classification of timing of injury of CP based on CT scan abnormalities (n = 626)*

Reference	Class	N	Type of CP	% Prenatal	% Perinatal	% Postnatal	% Unclassifiable
Wiklund et al., 1991 ⁶	I	83	H	44	39	0	17
Wiklund et al., 1991 ⁷	I	28	H	64	82	0	7
Chen, 1981 ⁹	III	282	M	33	54	13	0
Kolawole et al., 1989 ¹⁰	III	120	M	16	35	26	28
Taudorf et al., 1984 ¹¹	III	83	M	11	74	15	0
Molteni et al., 1987 ¹⁴	III	30	H	53	47	0	0
Total		626					
Mean values				32	50	12	6

d. Percent of patients with other etiologies of CP based on CT and clinical data (n = 702)*

Metabolic	4	Brain malformation	7
Genetic	2	Treatable condition	5

* Etiologies could not be determined in all patients in the studies listed in section a and in some patients more than one category of etiology was given. Data from the studies listed in section a.

H = hemiplegic; A = ataxic; M = mixed.

on physical examination (e.g., cataracts, chorioretinitis, dysmorphic features, myoclonus, dystonia) may suggest a specific etiology or an alternative diagnosis. An abnormality on MRI does not necessarily mean that the etiology of the motor deficit has been established. For example, the findings of diffuse cortical atrophy, delayed myelination, or polymicrogyria are nonspecific and only suggest that a CNS disturbance was present and may not allow one to know the underlying cause without a more specific workup.

CT. Data from 782 children with CP who had CT scans found abnormalities in 77% (range 62% to 93%) (see table 3a). For the class I studies (n = 140), 86% of children had abnormal scans whereas 78% of patients in class III studies (n = 642) had abnormal scans (there were no class II studies). The yield from CT scans varied depending on the type of CP (hemiplegic > ataxic > mixed > diplegic > quadriplegic > hypotonic > dyskinetic) with the percent abnormal in those with dyskinetic CP being much lower than in other forms of CP (table 3b).

Table 4 MRI in children with cerebral palsy (CP)

a. Overall yield of finding an abnormal MRI scan in children with CP

Reference	Class	N	Ages, y	Type of CP	% Abnormal
Krageloh-Mann et al., 1995 ¹⁵	I	56	5–17	SQ	91
Yin et al., 2000 ¹⁶	I	42	0.25–18	M	91
Candy et al., 1993 ¹⁷	II	22	0.25–2.25	M	77
Okumara et al., 1997, pt 2 ¹⁸	III	152	1–19	M	78
Cioni et al., 1999 ¹⁹	III	91	1–18.3	M	100
Jaw et al., 1998 ²⁰	III	86	0.33–13	M	95
Sugimoto et al., 1995 ²¹	III	70	0.75–15	M	100
Hayakawa et al., 1996 ²²	III	63	0.5–6	SD	79
Truwit et al., 1992 ²³	III	40	0.08–41	M	93
Yokochi et al., 1991 ²⁴	III	22	NA	D	68
Total no. of patients		644			
Mean abnormal MRI					89

b. Percent of patients with an abnormal MRI scan based on the type of CP (n = 286)

Mixed (n = 4)	100	Ataxic (n = 8)	75
Quadriplegic (n = 105)	98	Dyskinetic (n = 10)	70
Hemiplegic (n = 50)	96	Hypotonic (n = 6)	67
Diplegic (n = 102)	94		

c. Classification of timing of injury of CP based on MRI scan abnormalities*

Reference	Class	N	Type of CP	% Prenatal	% Perinatal	% Postnatal	% Unclassifiable
Krageloh-Mann et al., 1995 ¹⁵	I	56	SQ	14	39	0	46
Yin et al., 2000 ¹⁶	I	42	M	26	54	3	18
Cioni et al., 1999 ¹⁹	III	91	M	59	30	11	0
Jaw et al., 1998 ²⁰	III	86	M	29	28	3	8
Sugimoto et al., 1995 ²¹	III	70	M	44	37	0	19
Total		345					
Mean values				37	35	4	15

d. Percent of patients with other etiologies of CP based on MRI and clinical data (n = 682)*

Metabolic	0	Brain malformation	12
Genetic	1.4	Treatable condition	0.1

* Etiologies could not be determined in all patients in the studies listed in section a and in some patients more than one category of etiology was given. In some studies there were no data provided as to a specific etiology.

SQ = spastic quadriplegic; M = mixed; SD = spastic diplegic; NA = not available; D = dyskinetic.

When abnormal, CT scans are helpful in delineating the timing of the etiology of CP in most children (table 3c). For the class I studies (n = 111), onset was prenatal in 44%, perinatal in 43%, and postnatal in 0%. For the class III studies (n = 515), onset was prenatal in 29%, perinatal in 52%, and postnatal in 15%. In the remaining patients, the timing of the insult was not determined. Some of the more common etiologies of prenatal onset include intrauterine infection, stroke, toxemia, and placental abruption; of perinatal onset, etiologies include hypoxic ische-

mic encephalopathy, kernicterus, and trauma; and of postnatal onset, etiologies include infection, trauma, and progressive hydrocephalus. There was insufficient information from the CT studies to calculate an accurate breakdown by specific etiology for any of the three groups although in many of the individual cases a specific etiology was assigned. Etiologies tend to be different in term and preterm babies and are discussed further in the section on MRI.

A CT scan in a child with CP may on occasion detect conditions that are surgically treatable that

Table 5 MRI abnormalities in children with cerebral palsy based on preterm, term, and postnatal onset of insult

Abnormalities	Preterm (n = 335)	Term (n = 272)	Postnatal (n = 29)
Acquired lesions	261	178	22
PVL with other areas of injury	227	45	—
Diffuse encephalopathy (cortical/subcortical atrophy/ ventriculomegaly)	14	71	—
Focal ischemic/hemorrhagic (e.g., infarct porencephaly)	14	52	10
Multicystic encephalomalacia	3	10	—
Trauma (at birth or later)	0	0	4
Infection	3	0	8
Malformations*	48	55	0
Cortical dysplasia/polymicrogyria	8	18	—
Schizencephaly	6	11	—
Pachygyria/lissencephaly	5	9	—
Complex brain malformation	22	6	0
Agenesis/hypoplasia of the corpus callosum	3	3	—
Arachnoid cyst	1	0	—
Vermian/cerebellar hypoplasia	1	2	—
Hydrocephalus/holoprosencephaly/hydranencephaly	2	2	—
Miscellaneous/unknown	23	18	1
Miscellaneous etiologies	22	9	1
Delayed/abnormal myelination	1	9	0
Normal	3	21	6

Data from the following studies: Yin et al., 2000¹⁶; Okumara et al., 1997, pt 2¹⁸; Cioni et al., 1999¹⁹; Sugimoto et al., 1995²¹; Hayakawa et al., 1996²²; Krageloh-Mann et al., 1995¹⁵; Yokochi et al., 1991²⁴; Candy et al., 1993¹⁷; Jaw et al., 1998²⁰; Truwit et al., 1992.²³ Abnormalities considered insults on neuroimaging were designated as preterm (insult occurred before 38 weeks gestation whether infant born prematurely or at term), term (insult occurred after full term gestation in the perinatal period up to 1 month of age), or postnatal (insult occurred after 1 month of age in infants born prematurely or at term). Malformations were categorized as preterm if detected in infants born before 38 weeks gestation or term if detected in infants born after a full term pregnancy.

* The data in the malformations section of this table are separated into preterm, term, and post-term as that is how they were reported in the original reports. It is believed, however, that these malformations occur prenatally.

PVL = periventricular leukomalacia.

might not be detected by neurologic examination. The chance of finding a surgically treatable lesion based on data from 702 children with various types of CP is 5% (range 0% to 22.5%) (table 3d). This may vary as one class III study reported that 22.5% of 120 patients had potentially treatable lesions identified (hydrocephalus, porencephaly, arteriovenous malformation, subdural hematomas and hygromas, and a vermian tumor).¹⁰ The majority of other studies reported either no patients with potentially treatable lesions^{6,13,14} or lower incidences of 5%,⁹ 14%,¹¹ and 17%.⁸ On occasion, CT (as well as MRI) may detect abnormalities that suggest a potentially treatable inborn error of metabolism.

When the CT scan is abnormal, the presence of coexisting conditions should be anticipated. In studies of 240 children with CP, the presence of mental retardation was greater if scans were abnormal than if they were normal or had minor abnormalities (78% versus 39%).^{8,11-13} Three of these studies had additional data regarding the relation between CT findings and epilepsy.^{8,11,12} Ninety-one percent of children

with CP who had an abnormal CT scan had epilepsy or an abnormal EEG in contrast to only 38% of children whose scans were normal or minimally abnormal. One of these studies found normal EEG in 37%, paroxysmal EEG in 44%, and other abnormalities such as hemispheric asymmetry or suppression of activity in 19% of children.¹³ Although an abnormal CT suggested greater risk of coexistent conditions, the specific nature of these conditions (e.g., type of epilepsy) did not correlate with scan abnormalities.¹³

Conclusions. Data from three class I (75%) and six class III (77%) studies indicate that the yield of finding an abnormal CT scan in a child with CP is high (average of 77%) and related to the type of CP. Scan abnormalities may determine an etiology in many children but there were insufficient data to assess this further. Scan abnormalities may occasionally (i.e., 5 to 22%) identify treatable conditions and may suggest an increased risk for associated conditions such as mental retardation and epilepsy.

MRI. Data from studies involving 644 children with CP who had MRI scans found abnormalities in

89% (range 68% to 100%) (see table 4a). For the two class I studies (n = 98), 92% of children had abnormal scans whereas 77% of patients in one class II study (n = 22) had abnormal scans as did 89% of children in the class III studies (n = 524). The yield on MRI (table 4b) depended on the type of CP that was present (mixed > quadriplegic > hemiplegic > diplegic > ataxic > dyskinetic > hypotonic) and was somewhat different from that reported using CT.

MRI was helpful in determining whether the injury was prenatal, perinatal, or postnatal in onset (table 4c). Based on studies involving 345 children (class I n = 98; class III n = 247) with different types of CP, onset was prenatal in 37% (29% in class I and 45% in class III studies), perinatal in 35% (45% in class I and 31% in class III), and postnatal in 4% (1% in class I and 5% in class III studies). The yield from MRI also depended on whether the child with CP was born prematurely, at term, or whether CP was due to an insult later in life. Data from studies on 620 patients found that MRI scans were abnormal in 332/335 (99%) preterm infants, 251/272 (92%) term infants, and 23/29 (79%) infants older than 1 month (table 5). In most series, this was due to the fact that MRI was more sensitive in detecting periventricular leukomalacia, other perinatally acquired lesions, as well as subtle congenital anomalies of brain development. MRI can help define abnormalities based on the timing of insult in most patients as summarized in table 5. The etiology of CP in these children was suggested or established based on neuroimaging findings in combination with the clinical history.

Conclusions. Data from two class I (92%), one class II (77%), and seven class III studies (89%) indicate that the yield of finding an abnormal MRI scan in a child with CP is very high (average of 89%) and greater than that reported using CT (77%). The yield on MRI (as with CT) depends on the type of CP that is present. MRI is more likely to be abnormal in cases of CP associated with prematurity, showing abnormalities such as periventricular leukomalacia, compared to infants born at term. An etiology of CP can be determined in many patients based on the results of neuroimaging in combination with the clinical history.

Recommendations. 1. Neuroimaging is recommended in the evaluation of a child with CP if the etiology has not been established; for example, by perinatal imaging (Level A, class I and II evidence).

2. MRI, when available, is preferred to CT scanning because of the higher yield of suggesting an etiology and timing of insult leading to CP (Level A, class I–III evidence).

Metabolic and genetic testing. Should metabolic or genetic testing be routinely ordered in children with CP?

Evidence. Data from 2 class I, 13 class II, and 4 class III studies on 1,384 children with CP who underwent neuroimaging (CT or MRI) and who also had metabolic and genetic testing indicate that it is

rare to identify an underlying metabolic or genetic disorder. The mean incidence of metabolic (4%) and genetic disorders (2%) in those children who had CT scans was slightly higher than with MRI (metabolic, 0%; genetic, 1.4%) and did not vary substantially between the different classes of studies (see tables 3d and 4d). The low and variable incidence of metabolic and genetic disorders in these studies was, in part, due to patient selection factors, as in some studies children with metabolic disorders were excluded from analysis.¹⁷ Also, in these retrospective studies, the extent and type of metabolic or genetic testing was variable so that the exact prevalence of such disorders in children with CP remains uncertain.

Since the advent of neuroimaging, it has become more apparent that children with CP may have congenital brain malformations. Data from this same group of 1,426 children found that 7% of patients who had a CT scan and 12% of those who underwent MRI had major brain malformations such as lissencephaly, schizencephaly, or pachygyria (see tables 3d and 4d). Because these malformations are ever increasingly associated with specific genetic disorders (e.g., lissencephaly/Miller-Dieker syndrome/chromosome 17p13.3), their presence in affected children indicates the need for further genetic testing. Certain neurometabolic disorders (e.g., peroxisomal disorders such as Zellweger syndrome) may be associated with cerebral malformations and can present within the first years of life with a motor deficit that might appear to be nonprogressive. At present, no studies have prospectively evaluated children with CP with or without brain malformations to determine the incidence of metabolic or genetic abnormalities.

Metabolic disorders may on rare occasions masquerade as CP. Six small case series (i.e., class IV studies) describe 30 children who ultimately developed what appeared to be dyskinetic CP due to glutaric aciduria (type 1).^{25–30} These children typically develop normally until 5 to 10 months of age when they suffer an acute encephalopathy manifested by coma that is followed by dystonia, motor impairment, and macrocephaly (in about 60%).²⁵ Distinctive MRI and CT findings occur in half the patients and are manifested by frontal and temporal atrophy. Early diagnosis is important as glutaric aciduria is treatable; early intervention may prevent significant motor and cognitive impairment.²⁸ Other metabolic disorders presenting with symptoms suggestive of CP also have been reported in small case series and include Lesch-Nyhan syndrome,³¹ 3-methylglutaconic aciduria,³² pyruvate dehydrogenase deficiency,³³ argininemia,³⁴ cytochrome oxidase deficiency,³⁵ succinic semialdehyde dehydrogenase deficiency,³⁶ and female carriers of ornithine transcarbamylase deficiency.³⁷

Other childhood neurologic disorders (e.g., dopa-responsive dystonia, hereditary spastic paraplegia, ataxia telangiectasia) may initially be misdiagnosed as CP because of the slow rate of progression of symptoms. Other clinical or laboratory features of

Table 6 Associated conditions in children with cerebral palsy

Reference	Class	N	% Mental retardation	% Visual defects	% Speech-language disorders	% Hearing impaired
Zafeiriou et al., 1999 ⁴⁷	I	493	40	39	54	15
Murphy et al., 1993 ⁴⁸	I	204	65	10	NA	4
von Wendt et al., 1985 ⁴⁹	I	69	70	19	NA	7
Kolawole et al., 1989 ¹⁰	III	120	66	15	59	14
Total		886	52	28	38	12

NA = not available.

such conditions and observations over time that neurologic symptoms are progressive should suggest that the child does not have CP and mandates the need for further evaluation.

Conclusions. Metabolic or genetic causes for CP occur infrequently (i.e., 0 to 4%). However, the true incidence is unknown as there have been no prospective studies that have examined this issue. In almost all such cases, there are atypical complaints, features in the history of a progressive rather than a static encephalopathy, findings on neuroimaging that are representative of certain genetic or metabolic disorders, or a family history of childhood neurologic disorder with associated CP. Neuroimaging studies have shown that 7 to 11% of children with CP will have a brain malformation suggesting additional risk for genetic and possibly a metabolic etiology.

Recommendations. 1. Metabolic and genetic studies should not be routinely obtained in the evaluation of the child with CP (Level B, class II and III evidence).

2. If the clinical history or findings on neuroimaging do not determine a specific structural abnormality or if there are additional and atypical features in the history or clinical examination, metabolic and genetic testing should be considered (Level C, class III and IV).

3. Detection of a brain malformation in a child with CP warrants consideration of an underlying genetic or metabolic etiology (Level C, class III and IV evidence).

Coagulopathies. Should coagulation studies be performed in children with CP?

Evidence. Patients with hemiplegic CP frequently have suffered a prenatal or perinatal cerebral infarction. Data from three CT studies listed in table 3a (n = 196) found cerebrovascular occlusion, usually in the middle cerebral artery distribution, in 13%,⁶ 32%,¹¹ and 37%¹⁴ of individuals. There are insufficient data concerning the incidence of cerebral infarction or other causes of cerebrovascular compromise in children with other forms of CP.

Children, in contrast to adults, often have a coagulopathy, congenital heart disease, or an infectious process as the etiology of stroke.³⁸ However, none of the MRI series listed in table 4a focused on

children with hemiplegic CP. None of the reported series (see tables 3 and 4) included information indicating that a systematic examination for the etiology of infarction was performed. One class I study³⁹ and several class II studies have reported coagulation abnormalities as the etiology of neonatal cerebral infarction.³⁹⁻⁴² These have included Factor V Leiden deficiency, the presence of anticardiolipin or antiphospholipid antibodies, and Protein C or S deficiency. One class III study⁴² and several class IV case reports⁴³⁻⁴⁶ have also described the relation between neonatal cerebral infarction, coagulopathies, and a later diagnosis of hemiplegic CP.

Conclusions. Class I-III evidence indicates that cerebral infarction due to pre- or perinatal cerebrovascular occlusion occurs in 13 to 37% of children with hemiplegic CP. Class II and III evidence suggests that an etiology of cerebral infarction in this population may be due to a coagulation disorder and that the yield of testing will be higher if done in the neonatal period rather than if the child is evaluated later at the time of diagnosis of CP. There is insufficient evidence regarding the relation between coagulation disorders and other forms of CP.

Recommendations. Because the incidence of unexplained cerebral infarction seen with neuroimaging is high in children with hemiplegic CP, diagnostic testing for a coagulation disorder should be considered (Level B, class II-III evidence). There is insufficient evidence to be precise as to what studies should be ordered.

Associated conditions. What evaluations for associated conditions should be performed in children with CP?

Evidence. Children with CP often have associated conditions such as mental retardation or epilepsy that are equal in severity to their motor impairment. Because of the motor difficulties associated with CP, these conditions may not be readily recognized. These conditions may also seriously hamper the ability of a child to realize his or her intrinsic developmental potential.

Data from three class I and one class II studies involving 886 children with CP summarize the frequency of some of the major associated conditions that occur in children with CP (table 6).⁴⁷⁻⁴⁹ The incidence of mental retardation (52%), epilepsy (45%), ophthalmo-

Table 7 Prevalence of epilepsy in children with cerebral palsy

Reference	Class	N	Types of cerebral palsy	% Patients with epilepsy
Murphy et al., 1993 ⁴⁸	I	204	M	46
von Wendt et al., 1985 ⁴⁹	I	69	M	48
Miller and Cala, 1989 ⁸	I	29	A	59
Zafeiriou et al., 1999 ⁵⁰	II	493	M	36
Hadjipanayis et al., 1997 ⁵¹	II	323	M	42
Al-Sulaiman, 2001 ⁵²	II	151	M	54
Chambers et al., 1999 ⁵³	II	114	M	36
Bruck et al., 2001 ⁵⁴	II	100	M	62
Cioni et al., 1999 ¹⁹	II	91	M	35
Kwong et al., 1998 ⁵⁵	II	85	M	38
Kaushik et al., 1997 ⁵⁶	II	50	M	56
Taudorf et al., 1984 ¹¹	III	83	M	35
Senbil et al., 2002 ⁵⁷	III	74	M	42
Cohen and Duffner, 1981 ¹³	III	52	H	58
Total		1918		43

M = mixed; A = ataxic; H = hemiplegic.

logic defects (28%), speech and language disorders (38%), and hearing impairment (12%) are quite significant. Data from some of these studies also suggest that those children who have abnormal neuroimaging are more likely to have one or more of these deficits and in some of the studies severity of scan findings was associated with the severity of deficit.

Epilepsy. Should EEG be routinely performed in the assessment of children with CP?

Evidence. Given the higher frequency of epilepsy in children with CP, EEG is often considered during the initial evaluation.⁴⁷ The utility of EEG from a diagnostic perspective in this population has not been prospectively investigated. The vast majority of articles on EEG and CP are class III and IV studies that describe the frequency and types of seizures in children with different forms of CP but do not address the role of EEG in determining the etiology of CP nor in predicting the development of seizures in a child with CP.

Data from studies involving 1,918 children have found on average that 43% (range 35 to 62%) of children with CP develop epilepsy (table 7). In the three class I studies (n = 302), 48% had epilepsy in contrast to 42% of the children in the eight class II studies (n = 1,407) and 43% in the class III studies (n = 209). In none of these studies was there evidence that the EEG was useful in determining the etiology of the child's CP.

Data from one class II study compared patients with CP and epilepsy to those with epilepsy alone.⁵⁵ Children with CP had a higher incidence of epilepsy with onset within the first year of age (47% versus 10%), history of neonatal seizures (19% versus 3%), status epilepticus (16% versus 1.7%), need for polytherapy (25% versus 3%), and treatment with second-line antiepileptic drugs (31% versus 6.7%).

They also had a lower incidence of generalized seizures (28% versus 59%) and of remaining seizure free (37% versus 90%).⁵⁵ Factors associated with a seizure-free period of 1 year or more in epileptic children with CP include normal intelligence, single seizure type, monotherapy, and spastic diplegia. Similar findings have been observed by other investigators in the studies listed in table 7 and are summarized in the review by Wallace.⁴⁷ Children with CP who have abnormal neuroimaging studies are more likely to have epilepsy. One class I and two class II CT studies have examined the association between CT findings and epilepsy.^{8,11,13} Fifty-four percent of children with CP and an abnormal CT had epilepsy in contrast to only 27% of those who had a normal scan. In one study, EEG abnormalities (described as paroxysmal or asymmetric) were also much more commonly found in those children with an abnormal CT scan.¹³

The prevalence of epilepsy also varies depending on the type of CP that is present. Data from the studies listed in table 7 indicate that children with spastic quadriplegia (50 to 94%) or hemiplegia (30%) have a higher incidence of epilepsy than patients with diplegia or ataxic CP (16 to 27%). In patients with dyskinetic CP, it may occasionally be difficult to differentiate partial complex seizures from dyskinetic movements.

Conclusions. Although approximately 45% of children with CP develop epilepsy, in none of the retrospective studies involving 2,014 children was there evidence that the EEG was useful in determining the etiology of the child's CP. There is no evidence to make any recommendation regarding the child with CP who does not have a history of seizures as to whether an EEG should be ordered to screen for epileptiform abnormalities.

Recommendations. 1. An EEG should not be obtained for the purpose of determining the etiology of CP (Level A; class I and II evidence).

2. An EEG should be obtained when a child with CP has a history or examination features suggesting the presence of epilepsy or an epileptic syndrome (Level A; class I and II evidence).

Mental retardation. Cognitive and neuropsychological function in children with CP are commonly impaired.⁵⁸ In general, there is some but no absolute relation between the type of CP and severity of cognitive impairment. Children with spastic quadriplegia have greater degrees of mental impairment than children with spastic hemiplegia. Motor deficits of children with spastic CP appear to correlate with the severity of cognitive deficits in contrast to those children with dyskinetic CP where this relation is lacking.⁵⁸

Children with different forms of CP may be difficult to assess because of the motor deficits and in some forms of CP (e.g., spastic diplegia) the differences between performance and verbal intelligence test scores actually increase with age.⁵⁹ Laterality of hemiplegia may also be a contributing factor—those children with right hemiplegia may be more likely to have impaired language function due to left hemisphere injury,⁵⁹ although this remains controversial.⁶⁰ There is also a strong association between greater intellectual impairment in children with CP and the presence of epilepsy, an abnormal EEG, or an abnormal neuroimaging study.⁴⁷

Ophthalmologic impairments. Visual impairments and disorders of ocular motility are common (28%) in children with CP (see table 6). There is an increased presence of strabismus, amblyopia, nystagmus, optic atrophy, and refractive errors.⁶¹ Children whose CP is due to periventricular leukomalacia are also more likely to have visual perceptual problems. Many of these difficulties should be detected if currently accepted guidelines for vision screening in children with CP are employed.^{62,63}

Speech and language disorders. Because of bilateral corticobulbar dysfunction in many CP syndromes, anarthric or dysarthric speech and other impairments related to oral-motor dysfunction are common. For example, articulation disorders and impaired speech intelligibility are present in 38% of children with CP (see table 6).^{64,65} Because their impaired mobility can cause limited interaction with individuals in the environment, children with CP might not be able to develop the linguistic skills necessary to develop more complex speech patterns.⁶⁶ Language (as opposed to speech) deficits in CP go hand in hand with verbal intellectual limitations associated with mental retardation.⁶⁷ Oral-motor problems including feeding difficulties,⁶⁸⁻⁷⁰ swallowing dysfunction,^{65,70} and drooling⁷¹ may lead to potential serious impacts on nutrition and growth,⁷² oral health,^{73,74} respiration,⁷⁵ and self-esteem.

Hearing impairment. Hearing impairment occurs in approximately 12% of children with CP (see table 6).

This occurs more commonly if the etiology of CP is related to very low birth weight, kernicterus, neonatal meningitis, or severe hypoxic-ischemic insults. Children with CP who have mental retardation or abnormal neuroimaging studies are at greater risk for hearing impairment. Of concern are recent studies from the Centers for Disease Control that almost half the children found to have severe congenital hearing loss (with or without CP) in the greater Atlanta area were not recognized until almost age 3 years.⁷⁶ Established guidelines for neonatal audiometric screening have recently been published.⁷⁷

Conclusions. Children with CP are more likely to have associated conditions including mental retardation (52%), ophthalmologic defects (28%), hearing impairment (12%), and speech and language disorders (38%) and additional oral-motor deficits. Children with CP are also at greater risk for epilepsy (mean 43%; range 34 to 94%). However, there is no evidence that an EEG is helpful in determining the etiology of CP.

Recommendations. Because of the high incidence of associated conditions, children with CP should be screened for mental retardation, ophthalmologic and hearing impairments, and speech and language disorders (Level A, class I and II evidence). Nutrition, growth, and other aspects of swallowing dysfunction should be monitored. Further specific evaluations are warranted if screening suggests areas of impairment.

Recommendations for the evaluation of children with CP.

Although there is insufficient evidence to recommend the optimal sequence of tests to determine the etiology of CP, taking into account diagnostic yield and potential treatability, we propose the following consensus-based evaluation as outlined in the algorithm (figure). All children should undergo a detailed history and physical examination. It is important to determine that the child's condition is due to a static and not a progressive or degenerative neurologic disorder. It is also important to classify the type of CP as this has diagnostic implications as well as implications regarding associated problems. Because children with CP commonly have associated mental retardation, ophthalmologic abnormalities, hearing impairments, speech and language disorders, and disorders of oral-motor function, screening for these conditions should be part of the initial assessment. An EEG is recommended when there are features suggestive of epilepsy or a specific epileptic syndrome. In order to establish an etiology and prognosis in children with CP, neuroimaging is recommended, with MRI preferred to CT. However, if neuroimaging performed in the perinatal period provided an etiology of the child's CP, it may obviate the need for later study. Metabolic and genetic studies should not be routinely obtained in the evaluation of the child with CP. If the clinical history or findings on neuroimaging do not determine a specific structural abnormality or if there are additional and atypical features in

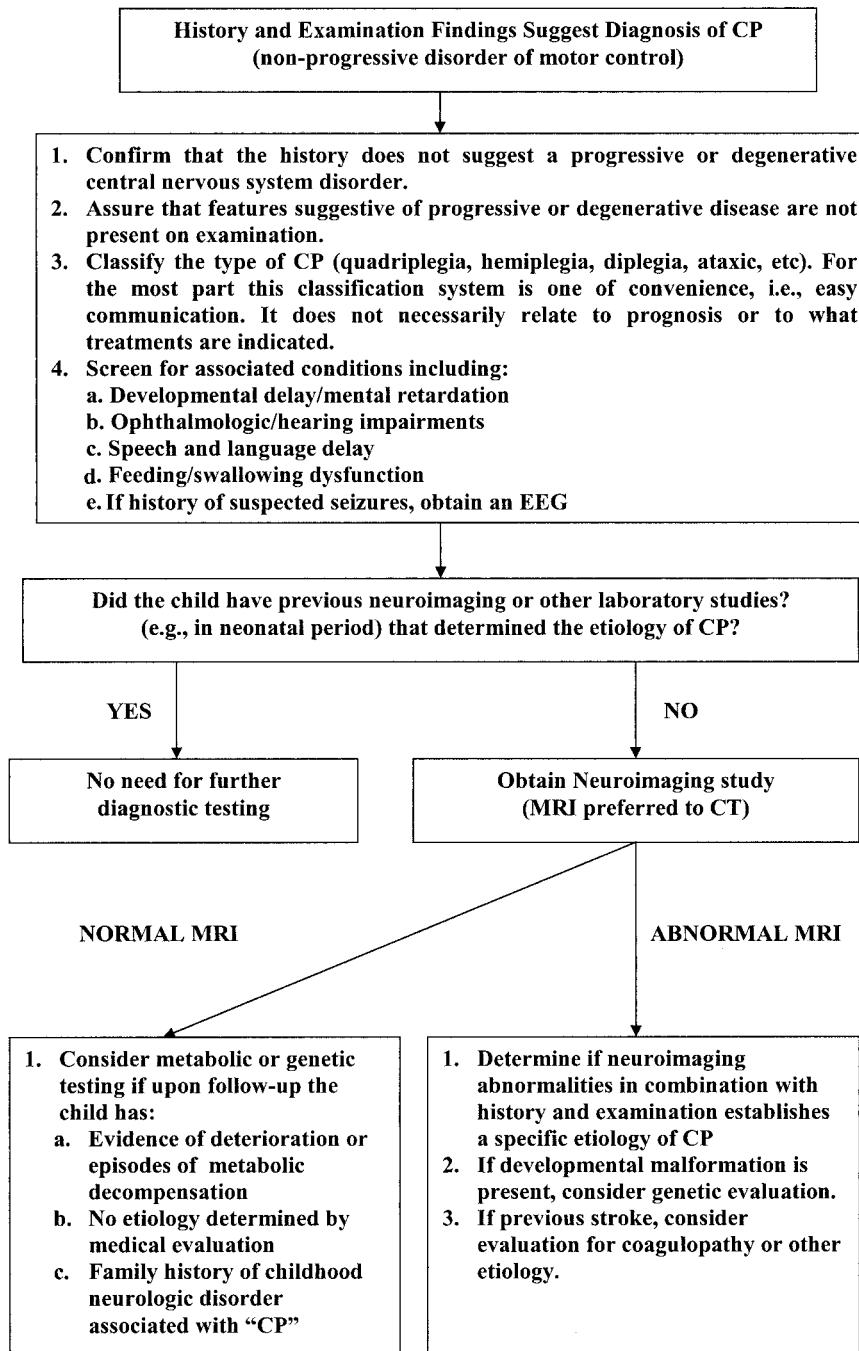


Figure. Algorithm for the evaluation of the child with cerebral palsy (CP). Screening for associated conditions (mental retardation, vision/hearing impairments, speech and language delays, oral motor dysfunction, and epilepsy) is recommended. Neuroimaging (MRI preferred to CT) is recommended for further evaluation if the etiology of the child's CP has not been previously determined. In some children, additional metabolic or genetic testing may be indicated.

the history or clinical examination, metabolic and genetic testing should be considered. Because the incidence of cerebral infarction is high in children with hemiplegic CP, diagnostic testing for a coagulation disorder should be considered.

Future research. 1. Prospective studies on the etiologic yields of genetic, metabolic, and neuroimaging diagnostic tests should be undertaken in large numbers of young children with CP compared with control subjects. This would permit the development of specific diagnostic algorithms.

2. Large prospective cohorts of children with CP should be studied to identify features based on CP

subtypes that can improve specific evaluation strategies and enhance etiologic yield.

3. It should be determined at what age and on what basis we can be certain that a child has CP sufficient to justify testing and at what age the yield will be optimal. Strategies of conducting testing simultaneously or sequentially need to be assessed. This should reduce unnecessary testing and provide cost-effective evaluations (i.e., a favorable balance between the cost of testing versus savings from early intervention, prevention of the birth of affected children).

4. Studies are needed to better characterize speech and language, ophthalmologic, auditory, oral-motor, nutrition, and growth deficits in children with

CP. Investigation of the sensorimotor impairments of children with CP is also needed so that studies of early intervention therapies might be done to improve the overall function of children who are likely to have multiple needs.

5. Issues related to quality of life and social support for families need further study. Included should be the benefits that medical testing confers by reducing parental concerns related to determining an etiology and by providing important information regarding prognosis, genetic counseling, and planning future educational and treatment needs.

6. Future research should also be directed to determine the underlying mechanisms causing CP that are associated with perinatal stroke, coagulopathies, genetic disorders, pre- and perinatal inflammatory diseases, and environmental factors.

Mission statement. The Quality Standards Subcommittee (QSS) of the AAN seeks to develop scientifically sound, clinically relevant practice parameters for the practice of neurology. Practice parameters are strategies for patient management that assist physicians in clinical decision making. A practice parameter is one or more specific recommendations based on analysis of evidence of a specific clinical problem. These might include diagnosis, symptoms, treatment, or procedure evaluation.

Disclaimer. This statement is provided as an educational service of the American Academy of Neurology and the Child Neurology Society. It is based on an assessment of current scientific and clinical information. It is not intended to include all possible proper methods of care for a particular neurologic problem or all legitimate criteria for choosing to use a specific procedure. Neither is it intended to exclude any reasonable alternative methodologies. The AAN and the CNS recognize that specific patient care decisions are the prerogative of the patient and the physician caring for the patient, based on all of the circumstances involved.

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Appendix 2: Members of the Practice Committee of the Child Neurology Society

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