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Garey H. Noritz, Nancy A. Murphy and NEUROMOTOR SCREENING EXPERT
PANEL

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CLINICAL REPORT

Motor Delays: Early Identification and Evaluation

abstract

FREE

Pediatricians often encounter children with delays of motor development in their clinical practices. Earlier identification of motor delays allows for timely referral for developmental interventions as well as diagnostic evaluations and treatment planning. A multidisciplinary expert panel developed an algorithm for the surveillance and screening of children for motor delays within the medical home, offering guidance for the initial workup and referral of the child with possible delays in motor development. Highlights of this clinical report include suggestions for formal developmental screening at the 9-, 18-, 30-, and 48-month well-child visits; approaches to the neurologic examination, with emphasis on the assessment of muscle tone; and initial diagnostic approaches for medical home providers. Use of diagnostic tests to evaluate children with motor delays are described, including brain MRI for children with high muscle tone, and measuring serum creatine kinase concentration of those with decreased muscle tone. The importance of pursuing diagnostic tests while concurrently referring patients to early intervention programs is emphasized. *Pediatrics* 2013;131:e2016–e2027

INTRODUCTION

The American Academy of Pediatrics (AAP) recommends developmental surveillance at all preventive care visits and standardized developmental screening of all children at ages 9, 18, and 30 months.¹ Recently, developmental screening instruments and their clinical interpretations have emphasized the early detection of delays in language and social development, responsive to rising prevalence rates of autism spectrum disorders in US children.² The most commonly used developmental screening instruments have not been validated on children with motor delays.^{3,4} Recognizing the equal importance of surveillance and screening for motor development in the medical home, this clinical report reviews the motor evaluation of children and offers guidelines to the pediatrician regarding an approach to children who demonstrate motor delays and variations in muscle tone. (This report is aimed at all pediatric primary care providers, including pediatricians, family physicians, nurse practitioners, and physician assistants. Generic terms, such as clinician and provider, are intended to encompass all pediatric primary care providers.)

RATIONALE

Gross motor development follows a predictable sequence, reflecting the functional head-to-toe maturation of the central nervous system.

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KEY WORDS

motor delays, development, screening, neurologic examination, early intervention

ABBREVIATIONS

AAP—American Academy of Pediatrics
CK—creatine phosphokinase
CPT—*Current Procedural Terminology*
DCD—developmental coordination disorder
DMD—Duchenne muscular dystrophy

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The guidance in this report does not indicate an exclusive course of treatment or serve as a standard of medical care. Variations, taking into account individual circumstances, may be appropriate.

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Although parents are reliable in reporting their child's gross motor development,^{5,6} it is up to the clinician to use the parent's report and his or her own observations to detect a possible motor delay.⁷

Gross motor delays are common and vary in severity and outcome. Some children with gross motor delays attain typical milestones at a later age. Other children have a permanent motor disability, such as cerebral palsy, which has a prevalence of 3.3 per 1000.⁸ Other children have developmental coordination disorder (DCD), which affects up to 6% of the population and generally becomes more evident when children enter kindergarten.⁹ When motor delays are pronounced and/or progressive, a specific neuromuscular disorder is more likely to be diagnosed. Motor delays may be the first or most obvious sign of a global developmental disorder. For infants, motor activities are manifestations of early development. It is often the case that children whose developmental trajectories are at risk may experience challenges in meeting early motor milestones. Establishing a specific diagnosis can inform prognosis, service planning, and monitoring for associated developmental and medical disorders. When the underlying etiology of motor delays is genetic, early recognition may assist parents with family planning. A timely diagnosis may reduce family stress related to diagnostic and prognostic uncertainties.⁵ For children with the few neuromuscular diseases for which treatments are available, outcomes may be improved when therapy is implemented early.¹⁰

Focus groups were conducted with 49 pediatricians at the AAP National Conference and Exhibition in 2010, and members of the AAP Quality Improvement Innovation Network were surveyed to ascertain current provider practices and needs regarding neuromotor screening.¹¹ Pediatricians described widely varying

approaches to motor examinations and identification of delays and expressed uncertainty regarding their ability to detect, diagnose, and manage motor delays in children. Participants requested more education, training, and standardization of the evaluation process, including an algorithm to guide clinical care (Fig 1).

THE ALGORITHM: IDENTIFYING CHILDREN WITH MOTOR DELAYS: AN ALGORITHM FOR SURVEILLANCE AND SCREENING

Step 1. Pediatric Patient at Preventive Care Visit

Each child's motor development should be addressed with other developmental and health topics at every pediatric preventive care visit.

Step 2. Is This a 9-, 18-, 30-, or 48-Month Visit?

All children should receive periodic developmental screening by using a standardized test, as recommended in the 2006 AAP policy statement "Identifying Infants and Children With Developmental Disorders in the Medical Home: An Algorithm for Developmental Surveillance and Screening."¹ Most children will demonstrate typical development without identifiable risks for potential delays. In the absence of established risk factors or parent or provider concerns, completion of a general developmental screening test is recommended at the 9-, 18-, and 30-month visits. These ages were selected, in part, on the basis of critical observations of motor skills development.

At the recommended screening visits, the following motor skills should be observed in the young child. These skills are typically acquired at earlier ages, and their absence at these ages signifies delay:

- 9-month visit: The infant should roll to both sides, sit well without support,

and demonstrate motor symmetry without established handedness. He or she should be grasping and transferring objects hand to hand.

- 18-month visit: The toddler should sit, stand, and walk independently. He or she should grasp and manipulate small objects. Mild motor delays undetected at the 9-month screening visit may be apparent at 18 months.
- 30-month visit: Most motor delays will have already been identified during previous visits. However, more subtle gross motor, fine motor, speech, and oral motor impairments may emerge at this visit. Progressive neuromuscular disorders may begin to emerge at this time and manifest as a loss of previously attained gross or fine motor skills.

An additional general screening test is recommended at the 48-month visit to identify problems in coordination, fine motor, and graphomotor skills before a child enters kindergarten.

- 48-month visit: The preschool-aged child should have early elementary school skills, with emerging fine motor, handwriting, gross motor, communication, and feeding abilities that promote participation with peers in group activities. Preschool or child care staff concerns about motor development should be addressed. Loss of skills should alert the examiner to the possibility of a progressive disorder.

Continuous developmental surveillance should also occur throughout childhood, with additional screenings performed whenever concerns are raised by parents, child health professionals, or others involved in the care of the child.

A summary of screening and surveillance for motor development based on the AAP "Recommendations for Preventive Pediatric Health Care" (also

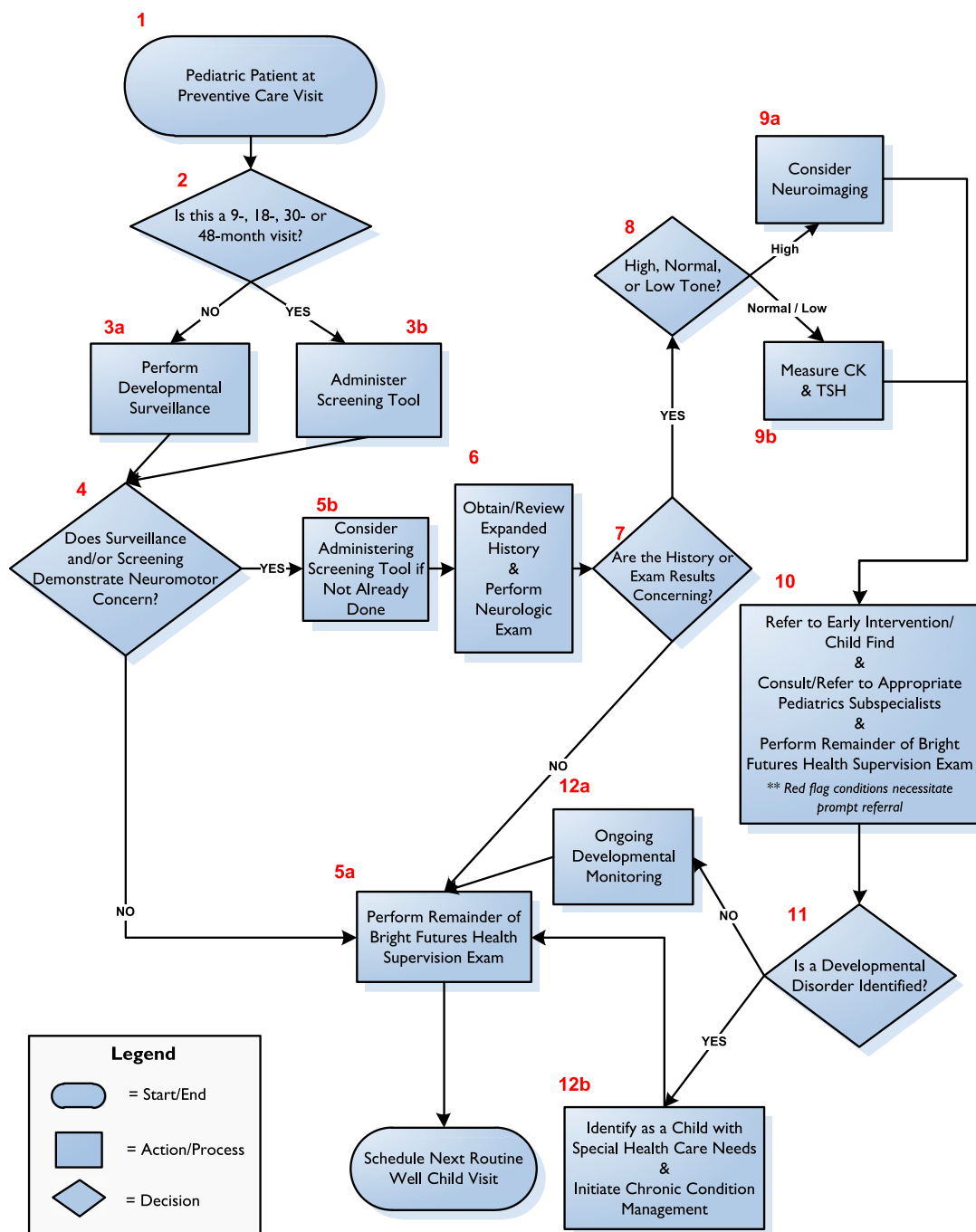


FIGURE 1
Identifying children with motor delays: an algorithm for surveillance and screening.

known as the periodicity schedule) is described in Table 1.¹² Listed are the mean ages at which typically developing children will achieve motor milestones. Marked delay beyond these ages warrants attention but does not necessarily signify a neuromotor disease.

Step 3a. Perform Developmental Surveillance

As the 2006 policy states, “Developmental surveillance is a flexible, longitudinal, continuous and cumulative process whereby knowledgeable health care professionals identify children who may have developmental problems.

Surveillance can be useful for determining appropriate referrals, providing patient education and family-centered care in support of healthy development, and monitoring the effects of developmental health promotion through early intervention and therapy.” The 5 components of

TABLE 1 Motor Milestones for Developmental Surveillance at Preventive Care Visits^a

Age	Gross Motor Milestones	Fine Motor Milestones
2 mo	Lifts head and chest in prone	
4 mo	Rolls over prone to supine; supports on elbows and wrists in prone	Hands unfisted; plays with fingers in midline; grasps object
6 mo	Rolls over supine to prone; sits without support	Reaches for cubes and transfers; rakes small object with 4 fingers
9 mo ^b	Pulls to stand; comes to sit from lying; crawls	Picks up small object with 3 fingers
1 y	Walks independently; stands	Puts 1 block in a cup; bangs 2 objects together; picks up small object with 2-finger pincer grasp
15 mo	Walks backward; runs	Scribbles in imitation; dumps small object from bottle, with demonstration
18 mo ^b	Walks up steps with hand held	Dumps small object from bottle spontaneously; tower of 2 cubes; scribbles spontaneously; puts 10 blocks in a cup
2 y	Rides on toy without pedals; jumps up	Builds tower and horizontal train with 3 blocks
2.5 y ^b	Begins to walk up steps alternating feet	Imitates horizontal and vertical lines; builds a train with a chimney with 4 blocks
3 y	Pedals; climbs on and off furniture	Copies a circle drawing; draws a person with head and one other body part; builds a bridge with 3 blocks
4 y	Climbs stairs without support; skips on 1 foot	Draw a person with 6 parts, simple cross; buttons medium-sized buttons

Adapted from Capute AJ, Shapiro BK, Palmer FB, Ross A, Wachtel RC. Normal gross motor development: the influences of race, sex and socioeconomic status. *Dev Med Child Neurol*. 1985;27(5):635–643; Accardo PJ, Capute AJ. *The Capute Scales: Cognitive Adaptive Test/Clinical Linguistic and Auditory Milestone Scale (CAT/CLAMS)*. Baltimore, MD: Paul H. Brookes; 2005; and Beery KE, Beery NA. *The Beery-Buktenica Developmental Test of Visual-Motor Integration (Beery VMI) Administration, Scoring and Teaching Manual*. Minneapolis, MN: NCS Pearson Inc; 2004.

^a These milestones generally represent mean age of performance of these skills.

^b It is recommended that a standardized developmental test be performed at these visits.

developmental surveillance are as follows: eliciting and attending to the parents' concerns about their child's development, documenting and maintaining a developmental history, making accurate observations of the child, identifying risk and protective factors, and maintaining an accurate record of documenting the process and findings.

A great breadth and depth of information is considered in comprehensive developmental surveillance. Much of this information, including prenatal, perinatal, and interval history will accumulate in the child's health record and should be reviewed at each screening visit.

Step 3b. Administer Screening Tool

Developmental screening involves the administration of a brief standardized tool that aids in the identification of children at risk for a developmental disorder. Many screening tools can be completed by parents and scored by nonphysician personnel; pediatric providers interpret the screening results. The aforementioned 2006 policy statement on developmental surveillance and screening provides a list of developmental screening tools and a discussion of how to choose an appropriate screening tool.

Step 4. Do Surveillance and/or Screening Demonstrate Neuromotor Concern?

Step 5a. Perform Remainder of Bright Futures Health Supervision Examination

Step 5b. Consider Administering Screening Tool if Not Already Done

The concerns of both parents and child health professionals should be included in determining whether surveillance suggests that the child may be at risk for developmental problems. If parents or health care providers express concern about the child's development, administration of a developmental screening tool to address the concern may be added.

Step 6. Obtain/Review Expanded History and Perform Neurologic Examination

Pediatricians can elicit key clinical information about a child's motor development from the child, parents, and family. Key elements are listed in Table 2. It is essential to ask parents broad, open-ended questions and listen carefully for any concerns. Some

concerns will be stated explicitly; others may be suggested through statements of perceived differences between a child's abilities and those of their age-matched peers. To broaden historical perspectives, clinicians can ask if extended family members, educators, or others who know the child well express any concerns about motor development. In instances of birth at earlier than 36 weeks' gestation, most experts recommend correcting for prematurity for at least the first 24 months of life.¹³ Last, while taking the history, clinicians should carefully watch the child's posture, play, and spontaneous motor function without the stressful demands of performance under deliberate observation. When children are tired or stressed, direct observation of motor skills may not be possible, and full reliance on historical information is needed.

Children with increased tone may attain motor milestones early, asymmetrically, or "out of order." These aberrant milestones may include rolling supine to prone before prone to supine, asymmetric propping with sitting, asymmetric grasp, development of handedness before 18 months,¹⁴ and standing before sitting.¹⁵

TABLE 2 Key Elements of the Motor History

Key Elements of Motor History	Example
Delayed acquisition of skill	Is there anything your child is <u>not</u> doing that you think he or she should be able to do?
Involuntary movements or coordination impairments	Is there anything your child <u>is</u> doing that you are concerned about?
Regression of skill	Is there anything your child <u>used</u> to be able to do that he or she can <u>no longer</u> do?
Strength, coordination, and endurance issues	Is there anything <u>other children</u> your child's age can do that are difficult for your child?

Physical Examination

The examination maneuvers described here are focused on medical home visits of children in the ambulatory setting. A discussion of newborn examination within the nursery setting is beyond the scope of this report; however, *Guidelines for Perinatal Care*, developed by the AAP Committee on Fetus and Newborn and American College of Obstetrics and Gynecology Committee on Obstetric Practice, provides further information.¹⁶

General Examination

When there are concerns regarding the quality or progression of a child's motor development, evaluation begins with a complete physical examination, with special attention to the neurologic examination and evaluation of vision and hearing. Children with motor delays related to systemic illness often show alterations in their level of interaction with their environment and general arousal. Careful assessments of head circumference, weight, and length/height with interpretation of percentiles according to Centers for Disease Control and Prevention or World Health Organization growth curves are essential and may facilitate early identification of children with microcephaly, macrocephaly, and growth impairments. Often, poor cooperation by the child may interfere with proper measurements, so any unexpected change in growth pattern should be rechecked by the clinician. Drooling or poor weight gain may suggest facial and oral motor weaknesses, and ptosis should prompt clinicians

to consider congenital myopathies or lower motor neuron disorders. Respiratory problems, such as tachypnea, retractions, and ineffective airway clearance, can accompany many neuromotor conditions. Careful palpation of the abdomen may reveal organomegaly suggesting glycogen storage diseases, sphingolipidoses, or mucopolysaccharidoses. The astute clinician can use findings from the general pediatric examination to individualize a diagnostic approach for a child with motor delays.

Neuromotor Examination

Ideally, children should be well rested and comfortable for neuromotor examinations. However, when toddlers and preschoolers are uncooperative, clinicians can still gain important diagnostic information by observing the quality and quantity of movement.

The cranial nerve examination includes eye movements, response to visual confrontation, and pupillary reactivity. Although fundoscopic examination may be difficult, red reflexes should be detectable and symmetric. The quality of eye opening and closure and facial expression, including smile and cry, should be observed. Oromotor movement can be observed and, in the older child formally tested, by observing palate and tongue movement and, if possible, by drinking through a straw or blowing kisses. Observation for tongue fasciculations and quality of shoulder shrug should be assessed. Strength is most easily assessed by functional observation. Attention to the quality and quantity of body posture and

movement includes antigravity movement in the infant and the sequential transition from tripod sitting with symmetrical posture to walking and then running, climbing, hopping, and skipping in the older child. Clinicians should note any use of a Gower maneuver, characterized by an ambulatory child's inability to rise from the floor without pulling or pushing up with his arms. Muscle bulk and texture, joint flexibility, and presence or absence of atrophy should be observed. Quality and intensity of grasp is most easily assessed by observation during play.

For the infant, postural tone is assessed by ventral suspension in the younger infant and truncal positioning when sitting and standing in the older infant.¹⁷ Extremity tone can be monitored during maturation by documenting the scarf sign in infants^{18,19} and popliteal angles after the first year (see Fig 2).²⁰ Persistence of primitive reflexes and asymmetry or absence of protective reflexes suggest neuromotor dysfunction. Unsteady gait or tremor can be a sign of muscle weakness. Diminution or absence of deep tendon reflexes can occur with lower motor neuron disorders, whereas increased reflexes and an abnormal plantar reflex can be signs of upper motor neuron dysfunction. Neuromotor dysfunction can be accompanied by sensory deficits and should be assessed by testing touch and pain sensation.

In older children, difficulties with sequential motor planning, or praxis, should be differentiated from strength and extrapyramidal problems. Dyspraxia refers to the inability to formulate, plan, and execute complex movements. Assessment includes the presence and quality of age-appropriate gross motor skills (stair climb, 1-foot stand, hop, run, skip, and throw) and fine motor skills (button, zip, snap, tie, cut, use objects, and draw). Many of these children also have hypotonia.²¹

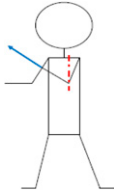
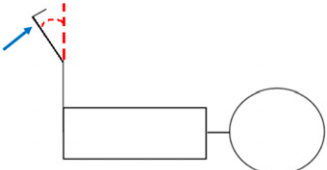
Physical Examination Maneuver		
	Scarf sign for tone assessment of shoulder girdle	Popliteal angle for hamstring tone assessment
Description	With child supine or sitting (if able), pull arm across to the opposite shoulder until resistance occurs. Note elbow position in relation to the bilateral midclavicular line and the midline of the chest.	With child supine, flex the hip to 90°, with child's torso supine on the table. Slowly extend leg at knee until resistance occurs. Note the angle between the lower leg and vertical.
IMPLICATIONS	FINDINGS^a	FINDINGS
Suspect normal tone	The elbow position is between the bilateral midclavicular lines	6° in children 1–3 y; 17°–27° in children 4 y, and 26° in children ≥5 y
Suspect low tone	The elbow crosses the midline to the contralateral midclavicular line	<6° in children ≥1 y
Suspect high tone	The elbow does not cross the ipsilateral midclavicular line	≥50° in children ≥1 y

FIGURE 2

Examples of physical examination maneuvers. Adapted from Mercuri E, Haataja L, Dubowitz L. Neurological assessment in normal young infants. In: Cioni C, Mercuri E, eds. *Neurological Assessment in the First Two Years of Life*. London, UK: Mac Keith Press; 2007:30–31. ^a In term infants, these findings do not change significantly after 3 months of age.

Step 7. Are the History or Examination Results Concerning?

After identifying concerns of motor development, primary care clinicians can perform key diagnostic tests. All testing should be performed in the context of the child's past medical history, including prenatal complications and exposures, perinatal problems, feeding, and growth. Family history is also important to identify any other relatives with developmental or motor issues, recurrent pregnancy loss, stillbirth, or infant death, which may lead to identification of an underlying genetic etiology. Findings on physical examination, such as unusual facial features or other known visceral anomalies, may suggest a specific genetic condition. The state-mandated newborn screening laboratory results should be reviewed, because normal results exclude many disorders and avoid unnecessary testing. Although newborn screening is

comprehensive, it does not test for all inborn biochemical disorders.

The algorithm (Fig 1) can be used to help guide appropriate initial testing. Table 3 lists “red flags” that should prompt the primary care pediatrician to expedite referral to diagnostic resources.

Step 8. High, Normal, or Low Tone?

Step 9a. Consider Neuroimaging

Increased tone in a child with neuro-motor delay suggests an upper motor neuron problem, such as cerebral palsy. The American Academy of Neurology recommends imaging of the brain, preferably by MRI, for patients suspected of having cerebral palsy.²² This test can be ordered within the medical home at the same time the patient is referred for specialist consultation for diagnosis.

Step 9b. Measure Creatine Phosphokinase and Thyroid-Stimulating Hormone Concentrations

When low to normal tone is identified, especially with concomitant weakness, investigations should target diseases of the lower motor neurons or muscles. Among the most common is Duchenne muscular dystrophy (DMD), characterized by weakness, calf hypertrophy, and sometimes cognitive or social delays. DMD usually presents at 2 to 4 years of age, but signs of weakness may be evident earlier. Becker muscular dystrophy is allelic to DMD but typically presents in older children and with a milder phenotype. Initial testing for all children with motor delay and low tone can be performed within the medical home by measuring the serum creatine phosphokinase (CK) concentration. The CK concentration is significantly elevated in DMD, usually >1000 U/L. As an X-linked disorder, there may be a family history of other affected male family members on the maternal side. However, DMD often presents in the absence of a family history for this disorder, with approximately one-third of cases being new mutations.²³ If the CK concentration is elevated, the diagnosis of DMD can usually be confirmed with molecular sequencing of the DMD gene. Other neuromuscular disorders include diseases of the peripheral motor nerves or muscles, such as myotonic dystrophy, spinal muscular atrophy, mitochondrial disorders, and congenital myasthenia gravis. Testing for these diseases should be performed by subspecialists, because these patients often require electrodiagnostic or specific genetic testing. Although congenital hypothyroidism will be identified by newborn screening, acquired hypothyroidism and hyperthyroidism can present in later infancy or childhood with motor delay

TABLE 3 “Red Flags” in the Evaluation of a Child With Neuromotor Delay

Red Flags: Indications for Prompt Referral	Implications
Elevated CK to greater than 3× normal values (boys and girls)	Muscle destruction, such as in DMD, Becker muscular dystrophy, other disorders of muscles
Fasciculations (most often but not exclusively seen in the tongue)	Lower motor neuron disorders (spinal muscular atrophy; risk of rapid deterioration in acute illness)
Facial dysmorphism, organomegaly, signs of heart failure, and early joint contractures	Glycogen storage diseases (mucopolysaccharidosis, Pompe disease may improve with early enzyme therapy)
Abnormalities on brain MRI	Neurosurgical consultation if hydrocephalus or another surgical condition is suspected
Respiratory insufficiency with generalized weakness	Neuromuscular disorders with high risk of respiratory failure during acute illness (consider inpatient evaluation)
Loss of motor milestones	Suggestive of neurodegenerative process
Motor delays present during minor acute illness	Mitochondrial myopathies often present during metabolic stress

and low to normal tone. It is reasonable to perform thyroid function studies (thyroxine [T4] and thyroid-stimulating hormone) as part of the general laboratory evaluation for children with low tone or neuromuscular weakness, even without classic signs of thyroid disease.

Cerebral palsy classically presents with spasticity, dystonia, or athetosis, but may also result in hypotonia. Children with cerebral palsy may have a history of perinatal insult with concomitant abnormalities on brain imaging. Other causes of hypotonia should be considered before the diagnosis of hypotonic cerebral palsy is given to a child with an uneventful perinatal history and normal brain imaging.

DCD may be present when a child's motor coordination performance is significantly below norms for age and intellect, unrelated to a definable medical condition that affects neuromotor function (such as cerebral palsy, ataxia, or myopathy). It can affect gait, handwriting, sports and academic participation, and self-help skills. More than half of individuals with DCD remain symptomatic through adolescence and young adulthood. Intervention, especially task-oriented approaches, can improve motor ability.⁸

Children with neuromotor abnormalities, who also have failure to thrive, growth abnormalities, dysmorphic facial features, or other visceral anomalies, may have a chromosome

abnormality, either common or rare. The American College of Medical Genetics and Genomics recommends microarray testing as the first-line chromosome study.²⁴ Because of the difficulty often encountered in interpretation of results, this test is typically ordered by a subspecialist familiar with this testing. Routine chromosome testing may be appropriate for children with weakness suspected as having recognizable disorders, such as Down syndrome (including mosaic Down syndrome), Turner syndrome, and Klinefelter syndrome. Fragile X syndrome is the most common inherited cause of cognitive impairment, and children with fragile X syndrome may have some element of motor delay. Genetic testing for fragile X syndrome should be considered in both boys and girls, whether they have dysmorphic facial features or a family history.

Common genetic conditions may present with early motor delays (Table 4). The 22q11.2 deletion syndrome (velocardiofacial syndrome) may present with hypotonia and feeding disorder in infancy and delayed motor milestones.²⁵ Noonan syndrome is also a common disorder, and although it is classically associated with short stature, webbed neck, ptosis, and pulmonary stenosis, the phenotype is highly variable, and developmental delays, especially motor delays, are common. Noonan

syndrome is genetically heterogeneous and may be caused by mutations in genes in the *ras* pathway.²⁶ Neurofibromatosis type 1, associated with mutations in the NF1 gene, can lead to developmental delays and hypotonia in infancy and early childhood. This condition should be suspected in children with hypotonia and multiple (greater than 6) café au lait spots.²⁷ Children with known or suspected genetic disorders may benefit from genetic consultation and genetic counseling for the family.

Step 10. Refer to Early Intervention/Child Find, and Consult/Refer to Appropriate Pediatric Subspecialists, and Perform Remainder of Bright Futures Health Supervision Examination

Observation

Mild abnormalities that are not accompanied by “red flag” findings (red flag conditions necessitate prompt referral) may be closely followed through “observation,” but a plan for new or worsening symptoms as well as a time-definite follow-up plan must be developed. Families should understand that clinical changes should prompt urgent reevaluation. This includes regression of motor skills, loss of strength, or any concerns with respiration or swallowing. This ensures that the progressive disorders are brought to medical attention immediately.

TABLE 4 Common Genetic Disorders for Which Neuromotor Delays May Be a Presenting Feature

Condition	Inheritance	Clinical Testing	Clinical Caveats
Angelman syndrome	Sporadic	Methylation testing for Prader-Willi/Angelman syndrome critical region, gene sequencing of UBE3A gene	Infantile hypotonia and delayed motor milestones, usually present with global delays; dysmorphic features are subtle in infancy.
Chromosome disorders	Many sporadic; high recurrence risk for unbalanced translocations if 1 parent has a balanced translocation	Chromosome analysis, single nucleotide polymorphism microarray	Some patients will have multiple anomalies and will have global developmental delays. Some may present in infancy or early childhood with delayed motor and/or speech milestones.
Down syndrome Klinefelter syndrome Rare deletions and duplications			Chromosome mosaicism also seen.
Deletion 22q11 syndrome (velocardiofacial syndrome)	Autosomal dominant (most cases new mutations)	Fluorescence in situ hybridization (FISH) for deletion 22q11.2	90% of cases new mutations. Feeding and speech disorders and cognitive impairment also seen. >50% will have a congenital heart defect.
DMD Becker muscular dystrophy	X-linked recessive	CK, sequencing of dystrophin gene	Becker and Duchenne muscular dystrophies are caused by mutations in different regions of the dystrophin gene. Becker muscular dystrophy has a later onset of symptoms with a less severe course; 67% of cases are inherited, 33% are new mutations.
Fragile X syndrome	X-linked	Gene sequencing and methylation analysis of FMR1 gene	Usually have global delays and cognitive impairment but may present in infancy or early childhood with predominantly motor delays. Males affected primarily, but females with FMR1 expansions may also be affected.
Mitochondrial myopathies	Autosomal recessive; X-linked recessive mitochondrial inheritance	Constitutional and mitochondrial genetic testing, lactate/pyruvate levels and ratio, serum amino acids	Genetic heterogeneity. May not present in infancy. Also at risk for cardiomyopathy, vision loss, hearing loss, cognitive disabilities.
Myotonic muscular dystrophy	Autosomal dominant	Gene sequencing for DMPK gene	May see anticipation with progression of phenotype in subsequent generations.
Neurofibromatosis type 1 (NF1)	Autosomal dominant	Usually a clinical diagnosis, gene sequencing NF1 gene	50% new mutations. Hypotonia most evident in infancy and early childhood. Suspect NF1 if hypotonia seen with multiple café au lait spots.
Noonan syndrome	Autosomal dominant	Gene sequencing for PTPN11 gene, genetically heterogeneous and multiple gene sequencing panels are available	Genetic heterogeneity. Commonly associated with short stature, ptosis, learning and developmental delays, hypotonia, pulmonary stenosis, cryptorchidism, cardiomyopathy.
Prader-Willi syndrome	Sporadic	DNA methylation testing for Prader-Willi/Angelman syndrome critical region	Hypogonadism, especially in boys. Hypotonia most evident in infancy and may be profound.
Spinal muscular atrophy, including congenital axonal neuropathy, Werdnig-Hoffmann disease, Kugelberg-Welander disease	Autosomal recessive	Gene deletion or truncation studies for SMN1 gene (95% to 98% of cases)	Usually presents in early infancy with severe hypotonia. Milder forms identified at later ages.

Depending on the nature of the suspected condition and the age of the child, it may be appropriate to have the child return to his or her medical home for a follow-up visit before the next Bright Futures health supervision visit. This will afford the opportunity

for an interval review of noted symptoms, new concerns, and changes in physical examination or other developmental findings.

Education with the family should not be overlooked or delayed, as a suspected condition can cause significant

anxiety.²⁸ Although the discussion may not be as in-depth as a situation in which diagnostic studies or referral is involved, families deserve a cogent and appropriate discussion of the findings that are being evaluated and what developmental trajectory is expected.

This may help assuage fears and increase compliance with follow-up plans.

Resources

All children with suspected neuromotor delay should be referred to early intervention or special education resources. Additionally, concurrent referrals should be made to physical and/or occupational therapists while diagnostic investigations are proceeding.²⁹ Even when a specific neuromotor diagnosis has not been identified, children with motor delays benefit from educationally and medically based therapies.

Each medical home must develop its own local resources and network of subspecialists for assistance with the diagnosis and management of young children with suspected motor delay. Depending on the setting, such subspecialists may include neurologists, developmental pediatricians, geneticists, physiatrists, or orthopedists. In some areas, availability of these resources may be limited, and waiting times may be long.³⁰ Direct physician-to-physician communication is recommended when red flags are identified (Table 3). Sharing digital photographs via a secure Internet connection may further expedite evaluations. However, the absence of red flags does not rule out the presence of significant neuromotor disease, and all children with motor delays should be thoroughly and serially evaluated.

Step 11. Is a Developmental Disorder Identified?

If a developmental disorder is identified, the child should be identified as a child with special health care needs, and chronic-condition management should be initiated (see Step 12b).

Step 12a. Ongoing Developmental Monitoring

If a developmental disorder is not identified through medical and developmental evaluation, the child should be

scheduled for an early return visit for further surveillance, as mentioned previously. More frequent visits, with particular attention paid to areas of concern, will facilitate prompt referrals for further evaluation when indicated.

Step 12b. Identify as a Child With Special Health Care Needs and Initiate Chronic Condition Management

When a child has delays of motor development, that child is identified as a child with special health care needs even if that child does not have a specific disease etiology. Children with special health care needs are defined by the Department of Health and Human Services, Health Resources and Services Administration, Maternal and Child Health Bureau as "...those who have or are at increased risk for a chronic physical, developmental, behavioral, or emotional condition and who also require health and related services of a type or amount beyond that required by children generally."³¹

Children with special health care needs benefit from chronic-condition management, coordination of care, and regular monitoring in the context of their medical homes. Primary care practices are encouraged to create and maintain a registry for the children in the practice who have special health care needs. The medical home provides a triad of key primary care services, including preventive care, acute illness management, and chronic-condition management. A program of chronic-condition management provides proactive care for children and youth with special health care needs, including condition-related office visits, written care plans, explicit comanagement with specialists, appropriate patient education, and effective information systems for monitoring and tracking. Management plans should be based on a comprehensive

needs assessment conducted with the family. Management plans should include relevant, measurable, and valid outcomes. These plans should be reviewed and updated regularly. The clinician should actively participate in all care-coordination activities for children with identified motor disorders. Evidence-based decisions regarding appropriate therapies and their scope and intensity should be determined in consultation with the child's family, therapists, pediatric medical subspecialists, and educators (including early intervention or school-based programs).

Children with established motor disorders often benefit from referral to community-based family-support services, such as respite care, parent-to-parent programs, and advocacy organizations. Some children may qualify for additional benefits, such as supplemental security income, public insurance, waiver programs, and state programs for children and youth with special health care needs (Title V). Parent organizations, such as Family Voices, and condition-specific associations can provide parents with information and support and can also provide an opportunity for advocacy.

RESOURCES

Internet resources are available (www.childmuscleweakness.org) for clinicians to view both typical and atypical motor findings. The identification of motor delays (or any chronic condition) in a child can trigger significant psychosocial stress for families.³² The effects of repeated medical visits, testing, and modifications to home and school environments can place a significant burden on even well-functioning families.³³ Appropriate psychological support should be implemented early. A consumer health librarian or medical librarian can be used by families to provide specific resources tailored to

individual needs (<http://www.nlm.nih.gov/medlineplus/libraries.html>).

For conditions with genetic basis or implications for family planning, medical genetics consultation and genetic counseling should be recommended. An international directory of genetics and prenatal diagnosis clinics can be found at <http://www.ncbi.nlm.nih.gov/sites/>

GeneTests/. Additional Web sites, such as www.rarediseases.org, offer information for both physicians and families.

Information on financial assistance programs should also be provided to families of children with established developmental disorders. They may qualify for benefits, such as supplemental

security income (<http://www.ssa.gov/pgm/ssi.htm>), public insurance (<http://www.medicaid.gov>), and Title V programs for children and youth with special health care needs (<http://internet.dscc.uic.edu/dsccroot/titlev.asp>). There also may be local community programs that can provide transportation and other assistance.

TABLE 5 CPT Codes for Developmental Screening

Services/Step in Algorithm	Notes	CPT Code	Comments
Pediatric preventive care visit	All preventive care visits should include developmental surveillance; screening is performed as needed or at periodic intervals	99381–99394 (EPSDT)	
Developmental/medical evaluation: Office or Other Outpatient Services Codes; New Patient	If performed by the physician as a new patient outpatient office visit	99201–99205	99204 is used for evaluations performed by the physician that are detailed and moderately complex or take at least 45 min (with more than half spent counseling); 99205 is used for evaluations that are comprehensive and highly complex or take 60 min (with more than half spent counseling)
Developmental/medical evaluation: Office or Other Outpatient Services Codes; Established Patient	If performed by the physician as an outpatient office visit	99210–99215	99214 is used for evaluations performed by the physician that are detailed and moderately complex or take at least 25 min (with more than half spent counseling); 99215 is used for evaluations that are comprehensive and highly complex or take 40 min (with more than half spent counseling)
Developmental/medical evaluation/Office or Other Outpatient Consultations Codes	If performed by the physician as an outpatient office visit	99241–99245	99244 is used for “moderate activities” of up to 60 min (with more than half spent counseling); 99245 is used for “high” activity of up to 80 min (with more than half spent counseling)
Developmental screening	Does not require any physician work; rather, clinical staff can score results and provide to physician for interpretation as part of E/M	96110	Reported in addition to E/M services provided on the same date
Developmental testing	Used for extended developmental testing typically provided by the medical provider (often up to 1 h), including the evaluation interpretation and report	96111	Reported in addition to E/M services provided on the same date
Identify as a child with special health care needs, and initiate chronic condition management	Children with special health care needs are likely to require expanded time and a higher level of medical decision-making found in these “higher-level” outpatient codes; these codes are appropriate for services in the office and for outpatient facility services for established patients; these codes may be reported using time alone as the factor if more than half of the reported time is spent in counseling	99211–99215	As above
Prolonged services	At any point during the algorithm when outpatient office or consultation codes are used, prolonged physician service codes may be reported in addition when visits require considerably more time than typical for the base code alone; both face-to-face and non-face-to-face codes are available in CPT	99354 99355 99358 99359	99354 for first 30–74 min of outpatient <u>face-to-face</u> prolonged services 99355 for each additional 30 min 99358 for first 30–74 min of <u>non-face-to-face</u> prolonged services 99359 for each additional 30 min

E/M, evaluation and management; EPSDT, Early and Periodic Screening, Diagnostic and Treatment program.

DEVELOPMENTAL SCREENING BILLING AND CODING

Separate *Current Procedural Terminology* (CPT) codes exist for developmental screening (96110: developmental screening) and testing (96111: developmental testing) when completing neuromotor screening and assessment. The relative values for these codes are published in the Medicare Resource-Based Relative Value Scale and reflect physician work, practice expenses, and professional liability expenses. Table 5 outlines the appropriate codes to use when billing for the processes described in the algorithm. Billing processes related to developmental screening and surveillance should be carefully reviewed to ensure that appropriate CPT codes are used to document screening procedures and ensure proper payment. CPT code 96110 does not include any payment for medical provider services. The expectation is that a nonphysician will administer the screening tool(s) to the parent and score the responses. The physician reviews and interprets the screening results; the physician's work is included in the evaluation and management code used for the child's visit. The preventive care (or new, consultative, or return visit) code is used with

the modifier 25 appended and 96110 listed for each screening tool administered. The CPT code 96111 includes medical provider work. This code would more appropriately be used when the medical provider observes the child performing a neuromotor task and demonstrating a specific developmental skill, using a standardized developmental tool.

CONCLUSIONS

The initial responsibility for identifying a child with motor delay rests with the medical home. By using the algorithm presented here, the medical home provider can begin the diagnostic process and make referrals as appropriate. Both during and after diagnosis, communication between the medical home and subspecialists is important,³⁴ and the medical home should remain fully engaged with the child's care as an integral part of chronic-condition management.

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