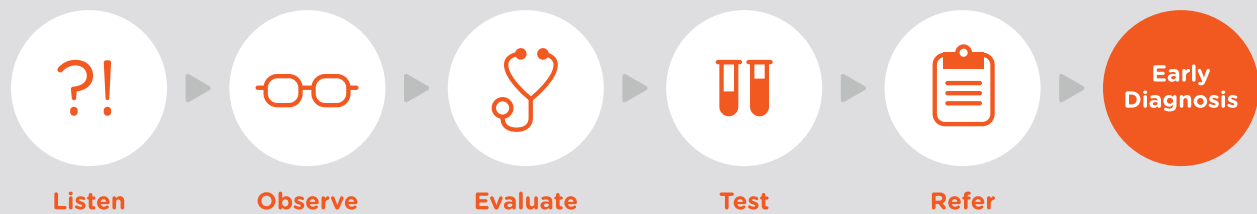


Early diagnosis makes a difference.

Learn the steps to identify pediatric muscle weakness and signs of neuromuscular disease.



Guide for therapists/specialists

National Task Force for Early Identification of Childhood Neuromuscular Disorders

Go to ChildMuscleWeakness.org for additional resources and video library.

Questions and comments to: info@ChildMuscleWeakness.org

Surveillance and Referral Aid

Target Audiences

- Therapists (PT, OT, Speech)
- Other early intervention professionals
- Physicians who see children referred from primary care for developmental delay, hypotonia, and other general, non-diagnostic indications, but are not themselves experts in neuromuscular disorders (such as rehabilitation medicine, developmental pediatricians, and orthopedics)

Goal

Among children (ages 6 months-5 years) referred for services/consultation based on developmental delays, **identify** those who might have weakness secondary to a neuromuscular disorder. **Listen** to parents' concerns about development. **Facilitate** early diagnosis of neuromuscular disorders, which is important because it helps target therapies and other interventions, improve outcomes, reduce family distress related to the diagnostic process, and provide the opportunity for accurate genetic counseling.

Use of the Surveillance Aid

- The guidance in this aid assumes that a therapist/specialist is following the child because he/she has an identified developmental delay and/or hypotonia.
- The guidance also assumes that the user understands normal motor development, and evaluates each child in light of his/her overall presentation and developmental history.
- The recommendations include communicating directly with the primary care provider. If direct communication is problematic, an alternative approach is to have the parents sign a release and send a copy of your report to the primary care provider.
- The materials that follow identify “red flags” by age, and suggest when **referral for diagnostic evaluation** should occur. In addition to the red flags, this aid includes a discussion of “yellow flags” that contribute to concerns about muscle weakness.
- This aid is designed for children who were born on or after 38 weeks of gestation. If a child is born prior to 38 weeks of gestation, please use adjusted age for developmental milestones.

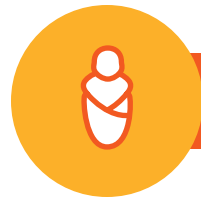
National Task Force for Early Identification of Childhood Neuromuscular Disorders

Go to ChildMuscleWeakness.org for additional resources and video library.

Questions and comments to: info@ChildMuscleWeakness.org

Signs of Weakness by Age

A motor surveillance aid for therapists, specialists, and other professionals who care for children, to aid in the identification of signs of motor weakness in children ages 6 months to 5 years.



Breathing (Infant+)

Description Look for sternal retraction, paradoxical breathing, abdominal breathing and accessory muscle use, nasal flaring, bell-shaped chest, and/or increased work to breathe.

Discussion None of these signs should be present, and the identification of these signs may indicate an urgent concern. Reasons for troubled breathing include (but are not limited to) neuromuscular disease, heart disease, and lung disease; some causes are transient and some are permanent.

Red Flag Any finding noted under the description is a red flag.

Response *Therapist*
Immediately contact the referring provider and request that the provider evaluate the child's breathing.

Specialist
Evaluate the need for urgent treatment/support and make immediate referral to specialists for further evaluation.

National Task Force for Early Identification of Childhood Neuromuscular Disorders

Go to ChildMuscleWeakness.org for additional resources and video library.

Questions and comments to: info@ChildMuscleWeakness.org



Pull to Sit (Infant+)

- Description** Evaluate pull to sit with attention to head lag, until achieved.
- Discussion** If a child has head lag at 4 months, carefully inspect neck flexors and evaluate other age-appropriate motor milestones (e.g., scores on standardized motor screening). Communicate concerns to family and referring provider and re-evaluate within one month.
- Tip** When evaluating pull to sit, consider decreased endurance of head control, such as decreased head control when carried, during eating, and when dressing.
- Yellow Flag** Head lag may present as torticollis.
- Red Flag** A child who has significant head lag at 5 months needs further evaluation.
- Response**
- Therapist*
Communicate specific concerns about head lag (in context of overall development and relevant environmental factors) to the family and referring provider. Encourage a referral to a specialist (consider pediatric neurology) for diagnostic evaluation.
- Specialist*
Draw a CK and refer for diagnostic workup. See *Motor Delay Algorithm* on page 12.

National Task Force for Early Identification of Childhood Neuromuscular Disorders

Go to ChildMuscleWeakness.org for additional resources and video library.

Questions and comments to: info@ChildMuscleWeakness.org



Sitting (6+ Months)

- Description** Evaluate sitting without support at 6 months and getting into sitting position at 9 months, or until achieved.
- Discussion** When evaluating sitting, look for continued progress through achievement of sitting skills: e.g., whether child can sit without using hands to prop self up; use both hands to hold a toy while sitting; maintain sitting when reaching for objects; and get in and out of sitting independently.
- Tip** Have a child reach for and hold a toy to evaluate ability to sit without propping. Assess the quality of the transition from supine to sitting (for example, rotating up to sit versus rolling to prone and pushing up to sit).
- Red Flag** A child who does not sit independently (i.e., un-propped and without requiring his/her hands for support) by 7 months **or** into a sitting position by 9 months needs further evaluation.
- Response**
- Therapist*
Communicate specific concerns about sitting (in context of overall development and relevant environmental factors) to the family and referring provider. Provide evidence for weakness rather than poor coordination as a cause for the delay. Encourage a referral to a specialist (consider pediatric neurology) for a diagnostic evaluation.
- Specialist*
Draw a CK and refer for diagnostic workup. See *Motor Delay Algorithm* on page 12.

National Task Force for Early Identification of Childhood Neuromuscular Disorders

Go to ChildMuscleWeakness.org for additional resources and video library.

Questions and comments to: info@ChildMuscleWeakness.org



Walking (12+ Months)

- Description** Observe each step of child's normal developmental progression to independent walking: pull up to stand, cruise holding on to furniture, stand alone, walk with one or both hands held, lower to sitting position from stand without falling; walk well.
- Discussion** Look at quality of posture and gait, in reference to how long child has been walking.
- Tip** Evaluate whether the child has problems walking when carrying an object the size of a small shoebox that is held at midline against the chest.
- Yellow Flag** Pronation of feet and/or ankles when walking; mild lumbar lordosis/ anterior pelvic tilt; increased width of base of support; lateral lean during stance (hip waddle); consistent toe walking.
- Red Flag** A child who does not walk well at 18 months, or shows regression in ability to walk, needs further evaluation.
- Response**
- Therapist*
Communicate specific concerns about walking (in context of overall development and relevant environmental factors) to the family and referring provider. Provide evidence for weakness rather than poor coordination as a cause for the delay. Encourage a referral to a specialist (consider pediatric neurology) for a diagnostic evaluation.
- Specialist*
Draw a CK and refer for diagnostic workup. See *Motor Delay Algorithm* on page 12.

National Task Force for Early Identification of Childhood Neuromuscular Disorders

Go to ChildMuscleWeakness.org for additional resources and video library.

Questions and comments to: info@ChildMuscleWeakness.org



Rise to Stand (12+ Months)

Description Watch for independent rise from floor from a supine position after child is able to walk well without assistance (generally 12-16 months); watch to see if child uses a Gower's maneuver (full or modified, by putting hands even briefly on knees or thighs) or cannot rise without pulling up.

Discussion Repeat any time concerns are raised about walking or other motor function, to evaluate for regression.

Tip An early Gower's can be very subtle.

Red Flag Child who cannot rise from floor to stand without support (including without using hands on knees or thighs to push up) by 18 months, or who shows regression in rise to stand, needs further evaluation.

Response *Therapist*
Communicate specific concerns about rise from floor (in context of overall development and relevant environmental factors) to the family and referring provider. Provide evidence for weakness rather than poor coordination as a cause for the delay. Encourage a referral to a specialist (consider pediatric neurology) for a diagnostic evaluation.

Specialist
Draw a CK and refer for diagnostic workup. See *Motor Delay Algorithm* on page 12.

National Task Force for Early Identification of Childhood Neuromuscular Disorders

Go to ChildMuscleWeakness.org for additional resources and video library.

Questions and comments to: info@ChildMuscleWeakness.org



Running (12+ Months)

Description Watch progression to run until running is achieved; watch for unusually frequent falls; ask about any concerns with walking, running, or frequent falls at all visits after milestone is achieved.

Discussion A child who does not run at 20 months: monitor closely and communicate concerns to the family and referring provider. Particularly note the quality of running, especially if there are other motor concerns.

Tip A true run means that both feet are off the floor simultaneously. If a child is attempting to run and is using a fast walk instead, his compensations will be exaggerated, e.g., he will pump/move arms for momentum and arch his back more, use a wider base of support, and will not be able to carry or drop objects while running.

Red Flag A child who does not run at 24 months, or shows regression in ability to run, needs further evaluation.

Response *Therapist*
Communicate specific concerns about running (in context of overall development and relevant environmental factors) to the family and referring provider. Provide evidence for weakness rather than poor coordination as a cause for the delay. Encourage a referral to a specialist (consider pediatric neurology) for a diagnostic evaluation.

Specialist
Draw a CK and refer for diagnostic workup. See *Motor Delay Algorithm* on page 12.

National Task Force for Early Identification of Childhood Neuromuscular Disorders

Go to ChildMuscleWeakness.org for additional resources and video library.

Questions and comments to: info@ChildMuscleWeakness.org



Jumping (12+ Months)

Description Watch child jump (straight up) starting at about 2 years. Look to see that both feet clear the ground. Watch child hop on one foot starting at about 3.5 years.

Red Flag Inability to jump with both feet simultaneously in the air by 2.5 years or inability to hop on one foot by 4 years requires further evaluation. Deterioration in these skills requires further evaluation.

Response *Therapist*
Evaluate in light of overall strength. Communicate specific concerns about jump and/or hop (in context of overall development and relevant environmental factors) to the family and referring provider. Provide evidence for weakness rather than poor coordination as a cause for the delay. Encourage a referral to a specialist (consider pediatric neurology) for a diagnostic evaluation if weakness is suspected or identified.

Specialist
Evaluate in light of overall strength. Refer for diagnostic workup (consider pediatric neurology) if weakness is suspected or identified. See *Motor Delay Algorithm* on page 12.

National Task Force for Early Identification of Childhood Neuromuscular Disorders

Go to ChildMuscleWeakness.org for additional resources and video library.

Questions and comments to: info@ChildMuscleWeakness.org



Climbing Stairs (12+ Months)

Description	Evaluate stair climbing at 2 years and up.
Discussion	Muscle weakness results in difficulty with stair climbing, which can be noted early. Children who are weak often avoid stairs. When going up stairs, they often crawl or stop to rest. Children who are weak use their arms excessively to pull themselves up the stair railing, and do not alternate feet (go step-over-step). Going down stairs, children who are weak also lead with one foot and usually have to hold onto the rail (but they do not have as much difficulty as when going up).
Red Flag	Any child who uses the stair railing to pull him/herself hand-over-hand up stairs at any age (not just using rail for balance) needs further evaluation. Any child who cannot go up stairs alternating feet step over step by 3.5 years needs further evaluation.
Response	<p><i>Therapist</i> Evaluate in light of overall strength. Communicate specific concerns about stair climbing (in context of overall development and relevant environmental factors) to the family and referring provider. Provide evidence for weakness rather than poor coordination as a cause for the delay. If weakness is suspected or identified, encourage a referral to a specialist (consider pediatric neurology) for a diagnostic evaluation.</p> <p><i>Specialist</i> Evaluate in light of overall strength. Draw CK and refer for diagnostic workup if weakness is suspected or identified. See <i>Motor Delay Algorithm</i> on page 12.</p>

National Task Force for Early Identification of Childhood Neuromuscular Disorders

Go to ChildMuscleWeakness.org for additional resources and video library.

Questions and comments to: info@ChildMuscleWeakness.org

Additional Red Flags

These additional red flags should prompt a diagnostic evaluation by a specialist, regardless of age:

- Tongue fasciculations
- Regression in developmental milestones
- CK level that is greater than 3x normal

Warning “Yellow Flags”

Yellow flags that should increase concern about muscle weakness (taking into account overall development):

- **Any developmental delay:** upon noting any developmental delay, evaluate motor development as part of the surveillance and screening
 - **Language delay:** note that neuromuscular disorders may present with language delays
- **Eating/feeding:** children who are very slow at swallowing, sucking, and/or chewing; frequent coughing during feeding; fatigue during feeding or stopping before satisfied; requiring more positional support than usual during feeding
- **Babbling:** if a child is not babbling by 10 months, evaluate motor development
- **Fatigue/decreased endurance:** tiring during routine tasks (e.g., eating, crawling, climbing, playing) can be an early sign of muscle weakness
- **Frequent falling:** frequent falls may be a sign of muscle weakness. Falls tend to be sudden, and are often characterized by collapsing straight down to the floor (not forward or over).

Developmental Norms

- pull to sit with no head lag*: 50% by 3.5 months, 75% by 4 months, 90% by 6.5 months
- sitting without support**: 50% by 5.9 months, 75% by 6.7 months, 90% by 7.5 months
- getting into sitting*: 50% by 8.5 months, 75% by 9.5 months, 90% by 10 months
- walking alone**: 50% by 12 months, 75% by 13.1 months, 90% by 14.4 months
- rise to stand from floor: corresponds with time of independent walking
- runs*: 50% by 16 months, 75% by 18.5 months, 90% by 21 months
- jump up*: 50% by 2 years, 75% by 2 years 3 months, 90% by 2.5 years
- hops*: 50% by 3.5 years, 75% by 4 years, 90% by 4 years 3 months

*Denver II **WHO Multicentre Growth Reference Study Group: WHO Motor Development Study

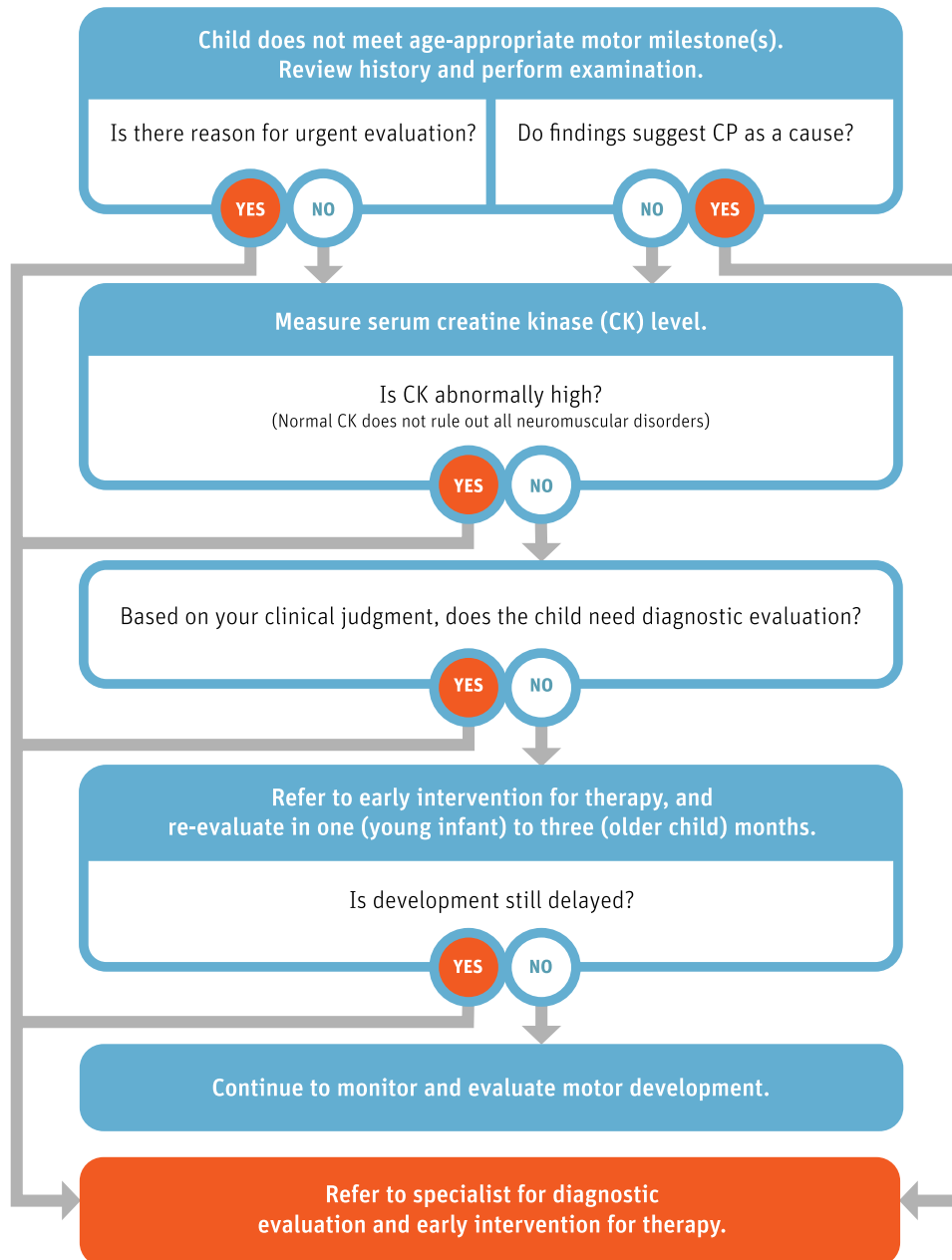
National Task Force for Early Identification of Childhood Neuromuscular Disorders

Go to ChildMuscleWeakness.org for additional resources and video library.

Questions and comments to: info@ChildMuscleWeakness.org

Motor Delay Algorithm

If a child does not meet age-appropriate motor development milestones, it may be a sign of a serious neurological or neuromuscular disorder. This chart will guide you through the appropriate tests and next steps for referral.



National Task Force for Early Identification of Childhood Neuromuscular Disorders

Go to ChildMuscleWeakness.org for additional resources and video library.

Questions and comments to: info@ChildMuscleWeakness.org