**MEASURE #3: Evaluation of Pulmonary Status Ordered**

**MUSCULAR DYSTROPHY**

**Measure Description**
All patients diagnosed with a muscular dystrophy (MD) who had a pulmonary status evaluation* ordered.

**Measure Components**

<table>
<thead>
<tr>
<th>Numerator Statement</th>
<th>Patients who had a pulmonary status evaluation* ordered.</th>
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<tbody>
<tr>
<td>*Pulmonary evaluation may include: referral for consultation with a pulmonologist, spirometry, maximal inspiratory pressure/maximum expiratory pressure (MIP/MEP), evaluation of cough and airway protection, screening for sleep disordered breathing</td>
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<thead>
<tr>
<th>Denominator Statement</th>
<th>All patients diagnosed with a muscular dystrophy.</th>
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</thead>
</table>

**Exceptions:**
- Medical exception for not ordering a pulmonary evaluation (i.e., patient cannot tolerate evaluation, MD phenotype does not warrant evaluation)
- Patient exception for not ordering a pulmonary evaluation (i.e., patient or family caregiver declines an evaluation)
- System exception for not ordering a pulmonary evaluation (i.e., clinic does not have the necessary equipment, patient cannot travel for testing, patient does not have insurance coverage)

**Supporting Guideline & Other References**
- Clinical assessment of respiratory health should be part of every medical consultation for children with neuromuscular weakness (NMW) and should be directed towards identifying progressive muscle weakness, ability to cope with respiratory infection, aspiration, progression of scoliosis and sleep-disordered breathing. [D]¹
- Vital capacity should be measured in all patients with neuromuscular disease who are capable of performing spirometry as part of the respiratory assessment. [C]¹
- C2. Clinicians should refer facioscapulohumeral muscular dystrophy (FSHD) patients with compromised pulmonary function studies (i.e., FVC < 60%) or with symptoms of excessive daytime somnolence or non-restorative sleep (e.g., frequent nocturnal arousals, morning headaches) for pulmonary or sleep medicine consultation for consideration of nocturnal sleep monitoring or nocturnal non-invasive ventilation to improve quality of life. (Level TBD)²
- G3. Clinicians should refer MD patients with excessive daytime somnolence, non-restorative sleep (e.g., frequent nocturnal arousals, morning headaches, excessive daytime fatigue), or respiratory insufficiency based on PFTs for pulmonary or sleep medicine consultation for consideration of non-invasive ventilation to improve quality of life. (Level B)²
- Obtain a detailed sleep history, evaluation of cough and airway protection, and serial measurements of FVC (sitting and supine) during routine office visits of patients with DM1 (Level A).³
- C1. Clinicians should obtain baseline pulmonary function tests on all patients with FSHD. Patients with abnormal baseline pulmonary function tests or with any combination of severe proximal weakness, kyphoscoliosis, wheelchair dependence, or co-morbid conditions that may affect ventilation (e.g., COPD, cardiac disease) should be monitored with pulmonary function testing at every clinic visit.²
- C3. All FSHD patients should have routine pulmonary function testing prior to all surgical procedures.²

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• G1. In MD patients at time of diagnosis, or if they develop pulmonary symptoms later in their course, clinicians should order pulmonary function testing (spirometry along with maximal inspiratory/expiratory force in both the upright and if normal, supine positions) or refer for pulmonary evaluation to identify and treat respiratory insufficiency. (Level B)4

• G1a. In patients with a known high risk of respiratory failure (limb girdle muscular dystrophy (LGMD2I & MFM), clinicians should obtain periodic pulmonary function testing (spirometry along with maximal inspiratory/expiratory force in the upright position, and if normal, in the supine position) or evaluation by a pulmonologist to identify and treat respiratory insufficiency. (Level B)4

• G2. It is not obligatory to refer patients with LGMD2B and LGMD2L for pulmonary evaluation or pulmonary function testing unless symptomatic. (Level C)4

• D1b. Pulmonary function should be monitored in the awake and sleep states on a regular basis. (Level B)4

• Each child with confirmed Duchenne muscular dystrophy (DMD) should undergo an evaluation of respiratory status early (between ages 4 and 6), and tests of respiratory function should be performed at every clinic visit thereafter.5

• Care by a pulmonologist should be increased to every 3 to 6 months after the initiation of assisted ventilation or an airway clearance device.5

• Objective evaluation at each clinic visit should include: oxyhemoglobin saturation by pulse oximetry, spirometric measurements of FVC, FEV1, and maximal mid-expiratory flow rate, maximum inspiratory and expiratory pressures, and peak cough flow.5

• Awake carbon dioxide tension should be evaluated at least annually in conjunction with spirometry. Where available, capnography is ideal for this purpose. Arterial blood gas analysis is not necessary for routine follow-up of patients with DMD. If capnography is not available, then a venous or capillary blood sample should be obtained to assess for the presence of alveolar hypoventilation.5

• Additional measures of pulmonary function and gas exchange may be useful, including lung volumes, assisted cough peak flow, and maximum insufflation capacity.5

• Carefully evaluate patients for evidence of other respiratory disorders, such as obstructive sleep apnea, oropharyngeal aspiration, gastroesophageal reflux, and asthma.5

• All children with abnormal overnight oximetry should undergo more detailed sleep monitoring with at least oxycapnography.1

• When there is doubt about the cause of sleep disordered breathing, overnight polysomnography or sleep polysomnography should be performed.1

• Obtain a detailed sleep history, evaluation of cough and airway protection, and serial measurements of FVC (sitting and supine) during routine office visits of patients with DM1. (Level A)3

• Perform an overnight sleep study in patients with clinical complaints suggestive of sleep-related respiratory dysfunctions. (Level C)3

• Carefully evaluate patients for evidence of other respiratory disorders, such as obstructive sleep apnea, oropharyngeal aspiration, gastroesophageal reflux, and asthma.5

• Review sleep quality and symptoms of sleep-disordered breathing at every patient encounter.6

• In areas where full polysomnography is not readily available, overnight pulse oximetry with continuous CO2 monitoring provides useful information about nighttime gas exchange, although sleep-disordered breathing not associated with desaturation or CO2 retention will not be detected. A simple capillary blood gas
upon arousal in the morning can demonstrate CO₂ retention, although not as sensitively as continuous capnography.⁶

- Assessment for sleep-disordered breathing should be carried out no less than annually for children with neuromuscular disease who have a vital capacity of <60% predicted and for children who have become non-ambulant because of progressive muscle weakness or who never attain the ability to walk. [D]¹
- In young children whose rate of disease progression is uncertain, or in older children who have shown a clinical deterioration or who are suffering with repeated infections, or who develop symptoms of sleep-disordered breathing, sleep assessment may need to be more frequent than once a year.¹


Rationale for the Measure

Some forms of MD are associated with oropharyngeal or ventilatory muscle weakness and those patients with these forms are at high risk for developing respiratory failure during the course of their disease. Patients with respiratory failure from neuromuscular-related weakness often do not have symptoms, such as dyspnea, that precede the onset of respiratory failure. Impending respiratory failure in these patients is often identified only with pulmonary function tests. Respiratory failure constitutes a major source of morbidity, interfering with daytime cognitive function and negatively affecting quality of life. Additionally, ventilatory and oropharyngeal weakness can threaten survival through the risk of upper airway obstruction and/or bellows failure.

Gap in care

A major contributor to morbidity and mortality in MD patients is respiratory failure. If not managed well and early on, it will bring adverse outcome. However, respiratory consultation does not take place in many patients with MD.¹ A Canadian report showed that in DMD, only 37% initially consulted respiratory therapists after a patient's first admission to hospital with respiratory complications.²

Opportunity for Improvement

Patients with respiratory failure secondary to muscle weakness often have improved quality of life with noninvasive pulmonary ventilation.³ Pulmonary function testing should therefore be done at regular intervals to identify the need for assistive respiratory equipment and initiate early noninvasive ventilation. Initiation of noninvasive ventilation can improve quality of life and prolong survival in patients with neuromuscular disease. Effective noninvasive strategies for management of hypoventilation, sleep-disordered breathing, and cough insufficiency are available for these patients.
A respiratory action plan should be enacted with increasing disease severity. Therapeutic measures comprise airway clearance, respiratory muscle training, noninvasive nocturnal ventilation, daytime noninvasive ventilation, and continuous invasive ventilation. At the advanced stage of respiratory failure, attention should be paid to complications related to long-term mechanical ventilation, such as pneumothorax and tracheal hemorrhage.

The American Thoracic Society (ATS) consensus statement on the respiratory care of patients with DMD has helped many patients receive improved care by offering clinicians guidance and helping medical directors of insurance companies make better decisions regarding use of technology to prevent morbidity and mortality. However, there is considerable work remaining to aid patients with DMD or types of MD with pulmonary complications.

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**AND**

CPT E/M Service Code:
99221, 99222, 99223 (Initial hospital care)
99231, 99232, 99233 (Subsequent hospital care)
99201, 99202, 99203, 99204, 99205 (Office or other outpatient visit-New Patient);
99211, 99212, 99213, 99214, 99215 (Office or other outpatient visit-Established Patient);
99241, 99242, 99243, 99244, 99245 (Office or Other Outpatient Consultation-New or Established Patient);
99304, 99305, 99306 (Initial nursing facility care, per day)
99307, 99308, 99309, 99310 (Subsequent nursing facility care, per day)
97001, 97002, 97003, 97004 (PT/OT evaluation)
99324, 99325, 99326, 99327, 99328 (Domiciliary visit, new patient)
99334, 99335, 99336, 99337 (Domiciliary visit, established patient)
99341, 99342, 99343, 99344, 99345 (Home visit, new patient)
99347, 99348, 99349, 99350 (Home visit, established patient)