American Academy of Neurology

Muscular Dystrophy

Quality Measurement Set

Status: For Public Comment Period
Public Comment Period: November 15-December 16, 2013
Physician Performance Measures (Measures) and related data specifications developed by the American Academy of Neurology (AAN) are intended to facilitate quality improvement activities by physicians.

These measures are intended to assist physicians in enhancing quality of care. Measures are designed for use by any physician who manages the care of a patient for a specific condition or for prevention. These Measures are not clinical guidelines and do not establish a standard of medical care, and have not been tested for all potential applications. The AAN encourages testing and evaluation of its Measures.

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American Academy of Neurology

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TOWARDS IMPROVING OUTCOMES FOR PATIENTS WITH A MUSCULAR DYSTROPHY

The American Academy of Neurology (AAN) formed a multi-disciplinary Muscular Dystrophy (MD) Measure Development Work Group to identify and define quality measures towards improving outcomes for patients with a muscular dystrophy. The majority of the available evidence that supported a gap in care focused on Duchenne muscular dystrophy, congenital muscular dystrophy, facioscapulohumeral muscular dystrophy and limb-girdle muscular dystrophy. Therefore this measurement set is predominantly focused on these types of muscular dystrophy.

The Work Group sought to develop measures to support the delivery of high quality care and to improve patient outcomes basing these measures on available clinical evidence focused on gaps in care in need of marked improvement. The Work Group considered the development of process, outcome, individual practitioner level and system level quality measures, where appropriate.

Importance of Topic

Prevalence and Incidence

- An estimated 1 of every 5,600 to 7,700 males 5 through 24 years of age had Duchenne/Becker Muscular Dystrophy (DBMD). That is approximately equal to a prevalence of 1.3 to 1.8 per 10,000 males 5 through 24 years of age in the four states. ([http://www.cdc.gov/mmwr/preview/mmwrhtml/mm5840a1.htm](http://www.cdc.gov/mmwr/preview/mmwrhtml/mm5840a1.htm))
- The most common form, Duchenne Muscular Dystrophy (DMD), affects 1 in every 3,500 to 6,000 male birthdays each year in the United States. DMD accounts for approximately 50 percent of all cases. ([Centers for Disease Control and Prevention, National Center on Birth Defects and Developmental Delays, July 17, 2013](http://www.cdc.gov/mmwr/preview/mmwrhtml/mm5840a1.htm))
- Becker Muscular Dystrophy (BMD)-affects males. Becker muscular dystrophy 1/18,000–1/31,000
- Congenital Muscular Dystrophy (CMD)-affects both males and females. 2.5/100,000
- Distal Muscular Dystrophy (DD): Most inherited in an autosomal dominant pattern but some follow a recessive pattern of inheritance. 1/10,000
- Emery-Dreifuss Muscular Dystrophy-primary affects males. Emery–Dreifuss muscular dystrophy 1/100,000
- Facioscapulohumeral Muscular Dystrophy (FSHD)-3rd most common form of MD 1/20,000.
- Myotonic dystrophy. Common form of MD. 1/8,000

Morbidity and Mortality

- Muscular dystrophy is associated with progressive muscle degeneration and weakness. Muscle weakness location depends upon the type of MD the patient has. It can affect the hips, pelvic area, thighs, shoulders, and skeletal (voluntary) muscles in the arms, legs, and truck. The heart and respiratory muscles can also be affected.
- Some types of MD shorter the person’s lifespan. People with DMD usually die of respiratory failure before they reach age 40. Life expectancy depends upon the degree of muscle weakness along with the presence of any cardiac or respiratory complications.
- Some types of MD are more severe and result in functional disability and loss of ambulation.
- Mortality/Morbidity Information by MD type ([National Institute of Neurological Disorders and Stroke and Muscular Dystrophy Association](http://www.ninds.nih.gov/disorders/muscular_dystrophy/muscular_dystrophy.htm)):
  - BMD-Survival is usually into old age.
o CMD-shortened life span
o DMD-life span ranges from 15 to 51. Muscle degeneration may be mild or severer.
o Distal MD-onset 20 to 60 years old. Progress is slow and not life threatening.
o Emery-Dreifuss-present in childhood and early teenage years with contractures. AT risk for stroke and sudden death from cardiac complications.
o FSHD-affects muscles of the factors, shoulders, and upper arms with progress weakness. Some affected individuals become severely disabled.
o Myotonic MD-delayed muscle relaxation, muscle wasting and weakness. Varies in severity and manifestations and affects many body systems in addition to skeletal muscles including the heart, endocrine organs, eyes, and gastrointestinal tract.
o Oculopharyngeal MD-onset 40 to 70 years old. Symptoms affect muscles of eyelids, face, and throat followed by pelvic and shoulder muscle weakness.

• There is no specific treatment currently available to stop or reverse any form of MD.

Health Related/Quality of Life
• Typically MD reduces the quality of life for those that are diagnosed with the disorder. Quality of life can be rated in terms of physical disability, pulmonary function, mobility, independence, etc.
• Health Related Quality of Life (HRQoL) of patient with MD using validated instruments in different age groups indicated that having MD negatively influences the HRQoL on several domains (physical symptoms, motor functioning, autonomy, cognitive functioning, social functioning, positive emotions and negative emotions). (Grootenhuis MA, De Boone, JD, Van Der Kooi A. Living with muscular dystrophy: health related quality of life consequences for adults and children. Open Access. Available at http://link.springer.com/article/10.1186%2F1477-7525-5-31#page-2)
• Overall, boys with DMD reported significantly lower QoL than their healthy peers. Despite decreased physical functioning, older boys seem to perceive better psychosocial QoL than perceived by their parents and by younger boys, unrelated to their need for mobility aids. http://pediatrics.aappublications.org/content/130/6/e1559
• Quality of life in DMD is not correlated with physical impairment or the need for noninvasive positive-pressure ventilation. The surprisingly high quality of life experienced by these severely disabled patients should be taken into consideration when therapeutic decisions are made. http://www.atsjournals.org/doi/full/10.1164/rccm.200503-322OC

Costs
• Medical costs are largely driven by outpatient care. Non-medical costs were driven by the necessity to move or adopt housing for the patient and paid caregiving. Annual per-patient costs for DMD $50,952 and $32,236 for Myotonic dystrophy. Population wide-national costs were $787 million (DMD) and $448 million (Myotonic dystrophy). (Larkindale J, Yang W, Hogan PF, et al. Cost of illness for neuromuscular disease in the U.S. Muscle & Nerve 2013. Accepted article. (Available at: mda.org/sites/default/files/Report_Summary-Cost_of_Illness.pdf )
• The yearly average cost in 2004 for medical care for privately insured individuals with any type of muscular dystrophy was $18,930, ranging from $13,464 at 5 through 9 years of age to $32,541 at 15 through 19 years of age. http://www.cdc.gov/ncbddd/musculardystrophy/data.html
• In 2005, the financial cost of MD was $435 million. Of this:
o $236.2 million (54.2%) was productivity lost due to lower employment, absenteeism and Premature death of Australians with MD;
o $117.8 million (27.1%) was the value of the informal care for people with MD, provided by parents and other close family or friends;
o $42.4 million (9.7%) was the deadweight loss from transfers including welfare payments (Mainly Disability Support Pension and Carer Payment) and taxation forgone;
o $29.7 million (6.8%) was other indirect costs such as aids and home modifications, formal care services, transport and the bring-forward of funeral costs; and
o $7.4 million (2.2%) was the direct health system expenditure.
o In per capita terms, this amounts to a financial cost of around $126,000 per person with
Gaps in Care and Opportunities for Improvement
The AAN worked with a medical librarian and did a supplementary web-based search to look for existing quality or performance measures for muscular dystrophy. Based upon the searches there are no existing quality measures for muscular dystrophies. There is a strong need for valid and reliable quality of care measures for muscular dystrophy disorder management. There measures are also needed on the health plan level.

Please see the individual measures listed for specific gaps in care and opportunities for improvement.

Disparities
- Muscular dystrophy occurs worldwide and affects all races.
- The mortality rate was higher in Whites than in Blacks, for both autosomal and X-linked MDs. The median age at death was lower in Blacks than Whites for both males and females. Cardiac complications were more commonly noted among MD-associated deaths in Blacks (38.9%) than Whites (28.6%). Respiratory infections were noted in about 20% of MD-associated deaths and were more common in winter than summer months. Potential reasons for the racial differences include differences in prevalence rates, rates of diagnosis, and reporting on death certificates. http://www.ncbi.nlm.nih.gov/pubmed/17022078

Rigorous Clinical Evidence Base
Clinical practice guidelines and peer-reviewed papers served as the foundation for the development of these performance measures. The majority of the available evidence that supported a gap in care focused on Duchenne muscular dystrophy, congenital muscular dystrophy, facioscapulohumeral muscular dystrophy and limb-girdle muscular dystrophy. Therefore this measurement set is predominantly focused on these types of muscular dystrophy.


Muscular Dystrophy Outcome Quality Measures
The work group discussed thoroughly many desired outcomes for the care of patients diagnosed with a muscular dystrophy. (See the draft list below under “Desired Outcomes for Patients with a Muscular Dystrophy”.) The Work Group drafted two outcome measures there were considered at the in-person work group member meeting.
- Quality of Life Patient Reported Outcome Measure for All Muscular Dystrophies
- MD Patient Satisfaction with Care Outcome Measure

However, these measures were all voted down primary because of the lack of strong guideline based high level of evidence recommendations or feasibility issues and thus these two measures were dropped from the draft measurement set.
**DESIRED OUTCOMES FOR PATIENTS WITH A MUSCULAR DYSTROPHY**

1. Quality of Life: Maintain or improve the patient reported quality of life.
   a. SF-36 scale commonly used.
      i. Measurements can be simpler scales, or even balanced Likert scales
      ii. Scales can be collected before a visit or periodically using on-line forms
   b. Limitations of overall quality of life combining multiple domains vs. single domain
      i. For example, measures of mood states, depression, anxiety

2. Independence
   a. Preserve or improve ambulation (focused on Duchenne Muscular Dystrophy)
      i. Measure timed walk
      ii. Capture age at time different ambulation aids are used (cane, walker, wheelchair, powered wheelchair)
   b. Social independence
      i. School attendance and participation
      ii. Employment
      iii. Participation in the community
      iv. Operate a motor vehicle?
   c. Maintain or improve function (ADLs)- PT/OT, exercise programs, orthoses
      i. Measure participation in exercise programs
      ii. Functional independence measures
   d. Communication
      i. Prevent, reduce social isolation (scales available)
   e. Maintain or improve nutrition
      i. Measure weight (gain/loss), for children height and weight gain, measure lean body mass

3. Health specific outcomes
   a. Appropriate diagnosis of the type of muscular dystrophy.
      i. Utilization of muscle biopsy
      ii. Utilization of genetic tests, when should they be used appropriately?
   b. Reduce cardiac morbidity and mortality
      i. Monitor for Congestive Heart Failure
      ii. Monitor for arrhythmia and blocks
   c. Reduce pulmonary complications and associated issues such as sleep disordered breathing (Notably in Limb Girdle Muscular Dystrophy)
      i. Compliance with CPAP, BiPAP
      ii. Compliance with other pulmonary care activities (e.g. use of in/exsufflator, suction, etc.)
      iii. Measure PFTs
   d. Reduce morbidity due to associated conditions in specific dystrophies (sleep apnea, cataracts, diabetes in DM1, retinopathy, deafness in FSHD, learning disabilities or cognitive dysfunction in DMD and some other dystrophies such as dystroglycanopathies etc.)
   e. Prevent bone loss
      i. Measure bone density, monitor for trend over time
   f. Prevent infections
      i. Following the recommended immunization schedule
      ii. Frequency of events

4. Improve care coordination, which is a very important aspect of management.
   a. Team approach.
      i. Proportion of patients in multidisciplinary MD clinics (MD-MDC) (real or virtual)
      ii. Proportion of people with MD with reasonable geographic and/or economic access to MD-MDC-system level
      iii. Proportion of people with MD seen in a MD-MDC with a care plan, or annual review of care plan

5. Increase patient and family engagement.
Do Not Cite. For Public Comment Period

a. This may include patient education,
   i. patient perception of adequacy of education
b. participation in advanced decision making,
   i. proportion of people with MD who have an advance care directive, who have expressed a desire to have an advanced care directive
c. active participation in treatment decisions, etc.
   i. Assessment of capacity of person with MD to consent to treatment decisions
   ii. use of shared decision making tools for people with MD
d. With children this may include involvement of the parent/guardian/care taker as a surrogate for the patient.

6. Increase patient satisfaction with care
   a. care satisfaction measure, a balanced Likert scale

7. Muscular Dystrophy Multidisciplinary Care Centers (MD-MDC):
   a. Understand if they provide better care than other type(s) of care settings. Requires a comparison group that can collect the same data, which may not be feasible except through patient entered registry data
   i. patient level outcomes: infection rate, survival, satisfaction, function (overlap with above outcomes)

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**Intended Audiences, Care Settings, and Patient Population**

The AAN encourages use of the measures by physicians and other health care professionals, where appropriate, to manage the care for patients with a muscular dystrophy. These measures are intended to be used to calculate performance or reporting at the practitioner level or system level. Performance measurement may not achieve the desired goal of improving patient care by itself. Measures have their greatest impact when they are used appropriately and are linked directly to operational steps that clinicians, patients, and health plans can apply in practice to improve care.
Do Not Cite. For Public Comment Period

### AAN Muscular Dystrophy Quality Measures QUALITY MEASURES

#### DMD Pharmaceutical Treatment

1. Patients with DMD Prescribed Appropriate Disease Modifying Pharmaceutical Therapy

#### MD Management

2. MD Multidisciplinary Care Plan Developed or Updated

3. Evaluation of Pulmonary Status Ordered

4. Evaluation of Cardiac Status Ordered

5. Scoliosis Evaluation Ordered

6. Referred for Physical, Occupational, or Speech/Swallowing Therapy

7. Nutrition or Growth Trajectory Status Monitored

8. Queried about Pain and Pain Interference with Function Evaluation

#### MD Planning and Patient Engagement

9. Counseled about advanced health care decision making, palliative care or end of life issues

These measures are discussed extensively in the latter half of this document with individual reviews and discussions of each measure.

### Institute of Medicine Domains of Health Care Quality

The landmark Institute of Medicine report *Crossing the Quality Chasm: A New Health System for the 21st Century* challenges all healthcare organizations to pursue six major aims of health care improvement: safety, timeliness, effectiveness, efficiency, equity, and patient centeredness. Please see below for how the work group feels these quality measures fit into the scope of these six major aims.

<table>
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<tr>
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<td>6. Referred for Physical, Occupational, or Speech/Swallowing Therapy</td>
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<td>7. Nutrition or Growth Trajectory Status Monitored</td>
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Other Potential Measures

The Work Group considered several other important constructs in MD care, though ultimately determined that the evidence was too weak, the gap in care was too small or the opportunity for improvement from the measure was too low to continue with the development of the measure. Thus, the draft measures were dropped and were considered not suitable for inclusion in this measurement set at this time.

Measure Harmonization

The AAN conducted an extensive literature search to seek out existing MD quality measures. There were no existing evidence-based MD quality measures by other measures developers found in the literature search, thus there was no need for harmonization.

Existing Quality Improvement (QI) Initiative or Collaborative for Measure Implementation

The American Academy of Neurology has developed a Performance in Practice program for Maintenance of Certification (MOC), NeuroPI (http://tools.aan.com/practice/pip/), which meets the American Board of Psychiatry and Neurology (ABPN) requirements for MOC Performance in Practice requirements. The NeuroPI will eventually contain a new module for Muscular Dystrophy based upon the measures developed in this measurement set.

Technical Specifications Overview

The AAN develops technical specifications for multiple data sources, including:

- Paper Medical Record/Retropective Data Collection Flow Sheet
- Electronic Health Record (EHR) Data
- Electronic Administrative Data (Claims)
- Expanded (multiple-source) Administrative Data

Administrative claims specifications are still being used for quality measure reporting to collect and report on quality measures. In the past the AAN has worked with the American Medical Association to create Current Procedural Terminology (CPT)-II codes to simplify the reporting burden. However, the AAN was notified in September 2013 that the AMA is no longer producing or supporting the development of CPT-II codes.

The AAN is in the process of creating code value sets and the logic required for electronic capture of the quality measures with EHRs. A listing of the quality data model elements, code value sets, and measure logic (through the CMS Measure Authoring Tool) for each of the MD measures will be made available at a later date.

Measure Exceptions and Exclusions

The AAN includes three possible types of exceptions or exclusions for reasons why a patient should not be included in a measure: medical, patient or system reasons.

- Medical exception examples:
  - not indicated (absence of organ/limb, already received/performed, other)
  - contraindicated (patient allergic history, potential adverse drug interaction, other)
- Patient exception examples:
  - patient declined
  - social or religious reasons
  - other patient reasons
- System exception examples:
  - resources to perform the services not available
  - insurance coverage/payer-related limitations
  - other reasons attributable to health care delivery system

For each measure, there must be a clear rationale to permit an exception or exclusion for a medical, patient, or system reason. For some measures, examples have been provided in the measure specification language of
instances that would constitute a reason why the patient should not be included in the measure. Examples are intended to guide clinicians and are not all-inclusive lists.

Although this methodology does not require the external reporting of more detailed exception or exclusion data, the AAN requests that physicians document the specific reasons for exception or exclusion in patients’ medical records for purposes of optimal patient management and audit-readiness. The AAN also advocates for the systematic review and analysis of each physician’s exceptions and exclusions data to identify practice patterns and opportunities for quality improvement. Please refer to measure specifications for each individual measure for information on the acceptable exceptions to be used for reporting each individual measure.

<table>
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<th>Testing and Implementation of the Measurement Set</th>
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<tr>
<td>The measures in the set are being made available without any prior testing. The AAN recognizes the importance of testing all of its measures and encourages testing of the MD measurement set for feasibility and reliability by organizations or individuals positioned to do so. The AAN welcomes the opportunity to promote the initial testing of these measures and to ensure that any results available from testing are used to refine the measures before implementation.</td>
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**DRAFT MEASURE #1: Patients with DMD Prescribed Appropriate Disease Modifying Pharmaceutical Therapy**

**MUSCULAR DYSTROPHY**

### Measure Description

All patients diagnosed with Duchenne muscular dystrophy (DMD) prescribed appropriate DMD disease modifying pharmaceutical therapy*.

### Measure Components

<table>
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<th>Numerator Statement</th>
<th>Patients prescribed appropriate DMD disease modifying pharmaceutical therapy*.</th>
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<td>*Current appropriate disease modifying pharmaceutical therapy for DMD: Corticosteroids</td>
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<th>Denominator Statement</th>
<th>All patients diagnosed with Duchenne muscular dystrophy (DMD).</th>
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#### Denominator Exceptions

- Medication exception for not prescribing disease modifying pharmaceutical therapy (eg medical contraindication; patient already on corticosteroid)
- Patient exception for not prescribing disease modifying pharmaceutical therapy (eg patient or family caregiver declines)
- System exception for not prescribing disease modifying pharmaceutical therapy (eg patient has no insurance to cover prescription and cannot afford it)

#### Supporting Guideline & Other References

- Treatment with corticosteroids to prevent the development or progression of scoliosis in DMD patients may be considered, even if the patient is wheelchair-bound.¹
- It is recommended to follow the Dutch guidelines [20] for the usage of corticosteroids in DMD patients.¹
- Benefits and side effects of corticosteroid therapy need to be monitored. Timed function tests, pulmonary function tests, and age at loss of independent ambulation are useful to assess benefits. An offer of treatment with corticosteroids should include a balanced discussion of potential risks. Potential side effects of corticosteroid therapy (weight gain, cushingoid appearance, cataracts, short stature (i.e., a decrease in linear growth), acne, excessive hair growth, gastrointestinal symptoms, and behavioral changes) also need to be assessed. If excessive weight gain occurs (20% over estimated normal weight for height over a 12-month period), based on available data, it is recommended that the dosage of prednisone be decreased (to 0.5 mg/kg/day with a further decrease after 3 to 4 months to 0.3 mg/kg/day if excessive weight gain continues) (Level A).²
- Deflazacort (0.9 mg/kg/day) can also be used for the treatment of DMD in countries in which it is available (Level A).²
- Prednisone has been demonstrated to have a beneficial effect on muscle strength and function in boys with DMD and should be offered (at a dose of 0.75 mg/kg/day) as treatment (Level A).²
- On the basis of this convincing literature, practice parameter guidelines, the panel strongly urges consideration of glucocorticosteroid therapy in all patients who have DMD. (Formal Consensus Statement)³
- Open discussion across the multidisciplinary team regarding the type and duration of specific interventions encourages transparency and shared decision-
Rationale for the Measure

Gap in Care

Duchenne muscular dystrophy (DMD) is a recessive X linked genetic disorder characterized by progressive muscle weakness and reduced muscle tone. Affecting only boys, it limits life expectancy to approximately 20 years. Care for patients with Duchenne muscular dystrophy (DMD) is poorly standardized. This leads to inequality in access to treatment.1

Although there is no cure, a Cochrane Review and AAN practice parameter concluded that prednisone may provide short term effective treatment that prolongs the ability to walk, reduces the complications such as scoliosis, respiratory insufficiency and cardiac impairment. Despite the well documented beneficial effects of corticosteroids in Duchenne muscular dystrophy, a population based study of corticosteroid use between 1991 and 2005 reported that only 50.9% of individuals had ever been on corticosteroids. The annual mean percent corticosteroid use varied widely from 8.4% to 80.2% across clinics.2 Another survey showed that nearly 10% of neuromuscular disease clinics do not offer such therapy.3

Glucocorticoids are currently the only medication available that slows the decline in muscle strength and function in DMD, which in turn reduces the risk of scoliosis and stabilizes pulmonary function.4 Approximately 16% of MDA Clinic directors report not using corticosteroids.3

Opportunity for Improvement

The goal of the use of glucocorticoids in the ambulatory child is the preservation of ambulation and the minimization of later respiratory, cardiac, and orthopaedic complications.3 Studies have shown that providing corticosteroid treatment early, such as in 2 to 4 year old DMD patients, can prolong the ability to walk, slow down respiratory decline and preserve left ventricular ejection fraction.5,6 There is also data to support the longer term (>3 years) use of corticosteroids to prolong ambulation, reduce the need for spinal stabilization surgery, improve cardiopulmonary function, delay the need for non-invasive ventilation, and improve quality of life and survival in patients with DMD.7

This quality measure has the opportunity to reduce the risk of scoliosis, stabilize pulmonary function, and potentially delay decline in respiratory and cardiac function.


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<td><strong>Measure purpose</strong></td>
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<td><strong>Data source</strong></td>
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<td>• Electronic health record (EHR) data</td>
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<tr>
<td>• Data registry</td>
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**Technical Specifications: Electronic Health Record/Registry (Under Development)**

Administrative claims data collection requires users to identify the eligible population (denominator) and numerator using codes recorded on claims or billing forms (electronic or paper). Unfortunately, DMD is not identifiable by an ICD-9 or ICD-10 code; rather it is grouped together with several other muscular dystrophy codes under one code (ICD-9: 359.1; ICD-10: G71.0 Muscular dystrophy). Therefore, the work group felt that this measure should be focused only on electronic health records and registries where the specific type of muscular dystrophy, DMD, can be easily identified. There is a SNOMED-CT code for Duchenne muscular dystrophy (disorder) Concept ID: 76670001 but this coding system is not commonly used for claims in the United States currently.

**Coding**

EHR or Registry diagnosis code of Duchenne Muscular Dystrophy. Full code value sets, logic and eMeasure HL7 format under development.

**Denominator**

SNOMED-CT Code for Duchenne muscular dystrophy (disorder) Concept ID: 76670001
## Measure Description

All patients diagnosed with a muscular dystrophy for whom a MD multi-disciplinary care plan* was developed, if not done previously, or the plan was updated at least once annually.

## Measure Components

| Numerator Statement | Patients for whom a MD multi-disciplinary care plan* was developed, if not done previously, or the plan was updated at least once annually. |

* MD Multi-disciplinary care plan should include a neurologist and access to the following clinicians as necessary depending on the specific MD and stage of the disease (listed in alphabetical order): advanced practice provider, cardiologist, dentist, dietitian, endocrinologist, gastroenterologist, genetic counselor, nurse practitioner, occupational therapist, orthopedic surgeon, palliative care specialist, pediatrician, physiatrist, physical therapist, physician assistant, primary care provider, psychiatrist, psychologist, pulmonologist, ophthalmologist, radiologist, respiratory therapist, sleep specialist, social worker, specialized nurse, speech/language pathologist, etc. |

| Denominator Statement | All patients diagnosed with a muscular dystrophy. |

| Denominator Exceptions | Exceptions:
- Medical reason for not developing or updating a multidisciplinary care plan (eg. plan was updated within 12 months of the date of the encounter)
- Patient reason for not developing or updating a multidisciplinary care plan (eg patient or family caregiver declines)
- System reason for not developing or reviewing a multidisciplinary care plan (eg patient has no insurance to cover the cost of a seeing specialists or other clinicians in a multidisciplinary care plan, cannot travel to see specialist, multidisciplinary services unavailable) |

| Supporting Guideline & Other References | Clinicians should refer patients with suspected muscular dystrophy to neuromuscular centers to optimize the diagnostic evaluation and subsequent management. (Level B)¹,²
- L1. Clinicians should refer patients with muscular dystrophy to a clinic that has access to multiple specialties (e.g., Physical Therapy, Occupational Therapy, Respiratory Therapy, Speech and swallowing Therapy, cardiology, pulmonology, orthopedics and genetics ) designed specifically to care for patients with muscular dystrophy and other neuromuscular disorders in order to provide efficient and effective long-term care. (Level B)¹
- AA1. Clinicians caring for children with CMD should consult a pediatric neuromuscular specialist for diagnosis and management. (Level B)²
- AA2. Pediatric neuromuscular specialists should coordinate the multidisciplinary care of CMD patients when such resources are accessible to interested families. (Level )²
- Coordination of clinical care is a crucial component of the management of DMD. This care is best provided in a multidisciplinary care setting in which the individual and family can access expertise for the required multisystem management of DMD in a collaborative effort. A coordinated clinical care role can be provided by a wide range of health-care professionals depending upon local services, including (but not limited to) neurologist or pediatric neurologists, rehabilitation specialists, |
neurogeneticists, pediatricians and primary-care physicians. It is crucial that the person responsible for the coordination of clinical care is aware of the available assessments, tools, and interventions to proactively manage all potential issues involving DMD. Includes: Diagnostics, Rehabilitation Management, Orthopedic Management, Psychosocial Management, Cardiac Management, Pulmonary Management, GI/Speech/Swallowing/Nutrition Management, and Corticosteroid Management. (Not a Guideline; Formal Consensus Process; No Level of evidence associated with recommendation)\textsuperscript{3,4}


Rationale for the Measure
A systematic review of muscular dystrophies has highlighted the medical complexity of caring for patients with muscular dystrophy. Such patients may develop cardiac, pulmonary, nutritional, and musculoskeletal complications that require the assistance of cardiologists, pulmonologists, orthopedists, physiatrists, physical therapists, occupational therapists, nutritionists, orthotists, and speech pathologists, in addition to neurologists. Additionally, myopathies with a limb-girdle, humeroperoneal, or distal pattern of weakness may be challenging to diagnose. A specific diagnosis provides patients with “closure,” assists genetic counseling, and directs monitoring for complications and optimal management.\textsuperscript{1}

Gap in Care
The purpose of having a multidisciplinary care plan is for patients with MD to enable the diagnosis of specific disorders, management of complications, optimize survival, and maintain quality of life. Such a plan has been recommended for the purpose of anticipatory care in patients with MD.\textsuperscript{2,4} The constitution of a multidisciplinary team is not standardized. The team often includes primary care providers, pulmonologists, cardiologists, ophthalmologists, physiotherapists, occupational therapy, orthopedists, physical medicine, orthopedics, neurologist and palliative care specialist. The needs of the patient may be different in the different forms of MD and at different stages of the disease, thus the requirement for specialists may change as well. One study indicated that Interdisciplinary Management of DMD should include the following: Diagnostics, rehabilitation management, orthopaedic management, psychosocial management, cardiac management, pulmonary management, GI/speech/swallowing/nutrition management, and corticosteroid management.\textsuperscript{2}

One study pointed out disparities in receipt of healthcare and related services in adult men with DBMD that can affect quality of life. These men only utilized half the services available...
to individuals with significant progressive conditions. Providers should be aware of low service utilization and focus on awareness and assistance to ensure access to available care.\(^5\)

Coordinated clinical care can bring awareness to potential issues and allow access to appropriate interventions that are critical for proper care in DMD. These include health maintenance and proper monitoring of disease progression and complications to provide anticipatory preventive care and optimum management.\(^4\)

**Opportunity for Improvement**

The implementation of multidisciplinary care plan should be early in the course of MD in order to achieve the best outcome in quality of life. To carry out such plan requires care coordination. Care coordination of all modalities of care (irrespective of whether the patient’s health is improving, remaining stable, or deteriorating) is essential. It should be orchestrated by a designated member of the team with whom the patient/family has direct contact. A nurse, nurse practitioner, or physician’s assistant is recommended. Such coordinator should be knowledgeable of the issues involved in muscular dystrophy and be capable of complex decision making. The coordinator may facilitate the implementation of anticipatory care, improve the knowledge base of disease-specific complications for the patient and treatment team and provide support to the clinic and patients. Early intervention may prevent joint contractures, scoliosis, foot and spine deformities, rigid spine, hip dislocation, and joint hyperextension.

This quality measure has the opportunity to increase the percentage of patients who have a multidisciplinary care and improve care coordination among specialists and other health care providers. If this measure is implemented in a registry, this quality measure could meaningfully increase care coordination and the overall care provided to patients with a muscular dystrophy.\(^5\)

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### Measure Designation

| Measure purpose | • Quality improvement  
| Type of measure | • Accountability  
| Level of Measurement | • Process  
|            | • Individual practitioner  

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Do Not Cite. For Public Comment Period

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<th>Care setting</th>
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<td>Inpatient Services</td>
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<td>Outpatient visits</td>
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<th>Data source</th>
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<td>Electronic health record (EHR) data</td>
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<td>Administrative Data/Claims Expanded (multiple-source)</td>
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<td></td>
<td>Paper medical record</td>
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**Technical Specifications: Administrative/Claims Data (Under Development)**

Administrative claims data collection requires users to identify the eligible population (denominator) and numerator using codes recorded on claims or billing forms (electronic or paper). Users report a rate based on all patients in a given practice for whom data are available and who meet the eligible population/denominator criteria.

The specifications listed below are those needed for performance calculation.

<table>
<thead>
<tr>
<th>Denominator (Eligible Population)</th>
<th>ICD-9 and ICD-10 Diagnosis Codes:</th>
<th></th>
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</thead>
<tbody>
<tr>
<td>359 Muscular dystrophies and other myopathies</td>
<td>ICD-9 Code</td>
<td>ICD-10 Code</td>
</tr>
<tr>
<td>359.0 Congenital hereditary muscular dystrophy</td>
<td>G71.2 Congenital myopathies</td>
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<tr>
<td>359.1 Hereditary progressive muscular dystrophy</td>
<td>G71.0 Muscular dystrophy</td>
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<tr>
<td>359.2 Myotonic disorders</td>
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<tr>
<td>359.21 Myotonic muscular dystrophy</td>
<td>G71.11 Myotonic muscular dystrophy</td>
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<tr>
<td>359.22 Myotonia congenita</td>
<td>G71.12 Myotonia congenital</td>
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<td>359.23 Myotonic chondrodystrophy</td>
<td>G71.13 Myotonic chondrodystrophy</td>
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<td>359.8 Other myopathies</td>
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<td>359.89 Other myopathies</td>
<td>G72.89 Other specified myopathies</td>
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<tr>
<td>359.9 Myopathy, unspecified</td>
<td>G72.9 Myopathy, unspecified</td>
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</table>

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

<table>
<thead>
<tr>
<th>Measure Components</th>
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<tbody>
<tr>
<td><strong>Numerator Statement</strong></td>
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<tr>
<td>Patients who had a pulmonary status evaluation* ordered.</td>
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<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, etc.</td>
</tr>
<tr>
<td><strong>Denominator Statement</strong></td>
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<tr>
<td>All patients diagnosed with a muscular dystrophy.</td>
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<tr>
<td><strong>Denominator Exceptions</strong></td>
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<tr>
<td>Exceptions:</td>
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<tr>
<td>• Medical exception for not ordering a pulmonary evaluation (eg. patient cannot tolerate evaluation, MD phenotype does not warrant evaluation)</td>
</tr>
<tr>
<td>• Patient exception for not ordering a pulmonary evaluation (eg patient or family caregiver declines an evaluation)</td>
</tr>
<tr>
<td>• System exception for not ordering a pulmonary evaluation (eg clinic does not have the necessary equipment, patient cannot travel for testing, patient does not have insurance coverage)</td>
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</table>

<table>
<thead>
<tr>
<th>Supporting Guideline &amp; Other References</th>
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<tbody>
<tr>
<td>• 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</td>
</tr>
<tr>
<td>• 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</td>
</tr>
<tr>
<td>• C2. Clinicians should refer FSHD patients with compromised pulmonary function studies (e.g. FVC &lt; 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</td>
</tr>
<tr>
<td>• G3. Clinicians should refer muscular dystrophy 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</td>
</tr>
<tr>
<td>• 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</td>
</tr>
<tr>
<td>• 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</td>
</tr>
<tr>
<td>• C3. All FSHD patients should have routine pulmonary function testing prior to all surgical procedures.²</td>
</tr>
<tr>
<td>• G1. In muscular dystrophy patients at time of diagnosis, or if they develop pulmonary symptoms later in their course, clinicians should order pulmonary function testing</td>
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</tbody>
</table>
spirometry along with maximal inspiratory/expiratory force in both the upright and 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 (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.** Clinicians might choose not 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 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 (3), maximum inspiratory and expiratory pressures, and peak cough flow (8).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. [O]1

- When there is doubt about the cause of sleep disordered breathing, overnight polysomnography or sleep polysomnography should be performed. [O]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.4

- 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]

- 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. [O]


Rationale for the Measure
Some forms of muscular dystrophy 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. A Canadian report showed that in DMD, only 37% initially consulted respirologists after a patient's first admission to hospital with respiratory complications.

Opportunity for Improvement
Patients with respiratory failure secondary to muscle weakness often have improved quality of
life with noninvasive pulmonary ventilation. 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 muscular dystrophy with pulmonary complications.

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99347, 99348, 99349, 99350 (Home visit, established patient)
**Measure Description**
Patients diagnosed with a muscular dystrophy who had a cardiac status evaluation* ordered.

**Measure Components**

<table>
<thead>
<tr>
<th>Numerator Statement</th>
<th>Patients who had a cardiac status evaluation ordered*.</th>
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<tr>
<td></td>
<td>*Cardiac evaluation may include: referral for a consultation with a cardiologist, electrocardiograms, echocardiograms, and other rhythm monitoring such as Holter monitoring, cardiac imaging, etc. that are relevant to the patient’s phenotype of muscular dystrophy.</td>
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<tr>
<th>Denominator Statement</th>
<th>All patients diagnosed with a muscular dystrophy.</th>
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**Denominator Exceptions**
- Medical exception for not ordering a cardiac evaluation (eg patient cannot tolerate the testing; MD phenotype is not associated with cardiac complications)
- Patient exception for not ordering a cardiac evaluation (eg patient or family caregiver declines)
- System reason for not ordering a cardiac evaluation (eg tests not available at the site, insurance does not cover evaluation)

**Supporting Guideline & Other References**
- The most useful risk factor for symptomatic cardiac disease in patients with myotonic dystrophy is the presence of asymptomatic EKG conduction abnormalities. The EKG should be used as an important screening test to determine the likelihood of cardiac complications (Level A).1
- C6. Clinicians do not need to obtain routine cardiac ECG or echocardiographic screening in FSHD patients either at diagnosis or during routine follow up.2
- C7. Clinicians should refer patients with FSHD for cardiac evaluation if they develop overt symptoms or signs of cardiac disease (e.g. shortness of breath, chest pain, palpitations).2
- E1a. If EKG or structural cardiac evaluation (e.g. echocardiography) are abnormal, or the patient has episodes of syncope, near-syncope, or palpitations, then rhythm evaluation (e.g., Holter monitor or event monitor) should be conducted to prevent cardiac morbidity and mortality. (Level B)3
- E4. Clinicians might choose not to refer patients with LGMD2A, LGMD2B, and LGMD2L for cardiac evaluation unless they develop overt cardiac signs or symptoms. (Level C)3
- E5. Clinicians should encourage female carriers of dystrophinopathy and emerinopathy to seek evaluation by a neuromuscular specialist and a cardiologist to assess for skeletal muscle and cardiac muscle involvement and to proactively treat cardiac involvement. (Level B)3
- Regular cardiac evaluations should start at school age and patients should be seen by a pulmonologist twice a year beginning at age 12 or when their FVC deteriorates to 80% of normal.4
- All individuals with DMD require regular cardiac evaluation with annual electrocardiograms and echocardiograms, starting at least by school age.5
- Cardiac care of the patient with DMD or BMD should begin after confirmation of the diagnosis. The patient should be referred for evaluation to a cardiac specialist.
with an interest in the management of cardiac dysfunction and/or neuromuscular disorders. (No Level of evidence listed).6

• A complete cardiac evaluation should include (but not be limited to) a history and physical examination, electrocardiogram, and transthoracic echocardiogram. Consideration should be given to a multi-gated acquisition study (MUGA) or cardiac MRI in patients with limited echocardiographic acoustic windows. (No Level of evidence listed).6

• Signs and symptoms of cardiac dysfunction should be treated. Consideration should be given to the use of diuretics, angiotensin-converting enzyme inhibitors, and/or β-blockers. (No Level of evidence listed).6

• Abnormalities of cardiac rhythm should be promptly investigated and treated. Periodic Holter monitoring should be considered for patients with demonstrated cardiac dysfunction. (No Level of evidence listed).6

• Patients with DMD should be routinely managed in early childhood with a complete cardiac evaluation at least biannually. (No Level of evidence listed).6

• For patients with DMD, yearly complete cardiac evaluations should begin at approximately 10 years of age or at the onset of cardiac signs and symptoms. However, individuals demonstrating these signs and symptoms are relatively late in their course. (No Level of evidence listed).6

• For patients with BMD, complete cardiac evaluations should begin at approximately 10 years of age or at the onset of signs and symptoms. Evaluations should continue at least biannually. (No Level of evidence listed).6


Rationale for the Measure
Many, though not all, dystrophy subtypes have associated cardiac involvement. There is an important risk of symptomatic involvement of both skeletal muscle and cardiac muscle in female carriers of dystrophinopathy and emerinopathy. About 15% of carriers of
dystrophinopathy have cardiac involvement before 15 years of age. This increases to about 45% in patients above 15 years of age. Similarly, about 18% of female carriers of emerinopathy over the age of 60 years have typical ECG abnormalities. Dystrophy patients or symptomatic carriers with cardiac involvement often do not have symptoms such as chest pain, pedal edema, or palpitations that precede cardiac morbidity or sudden cardiac death. Serious cardiac manifestations in patients with dystrophy are often identified only with cardiology testing. The detection and appropriate management of cardiac dysfunction are important to reduce morbidity and mortality. Patients with dystrophy often have improved quality of life following appropriate pharmacologic treatment, device placement, or surgical intervention for their cardiac involvement.1

Our systematic review found that dystrophy patients with certain genetic subtypes (LGMD2A, LGMD2B, and LGMD2L) are at very low risk of concomitant cardiac involvement during the course of their disease. Asymptomatic patients with these dystrophy subtypes would not benefit from cardiac testing. They would only be exposed to the added risk and costs associated with this testing. The quality of life in asymptomatic dystrophy patients with genetic subtypes at very low risk of concomitant cardiac involvement is not improved by cardiology evaluation and testing.1

Gap in care
Cardiac involvement occurs as a degenerative process with fibrosis and fatty replacement of the myocardium in many patients with MDs. Cardiac rhythm abnormalities are frequent and are a significant cause of morbidity and mortality for patients affected by Duchenne or Becker muscular dystrophy.2 Such changes cause dilated cardiomyopathy in DMD, BMD and LGMD, cardiac arrhythmias in myotonic dystrophy, Emery-Dreifuss, LGMD and FSHD. Therefore, timely evaluation of cardiac status is important to prevent sudden death due to arrhythmias, morbidity due to cardiomyopathy and resultant congestive heart failure and to improve outcome.

Cardiac evaluation is suboptimal even in female carries of DMD and BMD. One study showed that only 64.4% of the carriers had ever had a heart test; 18.3% had seen a cardiologist in the past year. Even when carriers informed their provider about the condition, only 70.2% had ever had a heart test and only 21.4% had seen a cardiologist in the past year.3

Opportunity for Improvement
Most DMD patients remain asymptomatic for years in spite of the progression of cardiac dysfunction because of their limited daily activities. Early detection of cardiac dysfunction and treat appropriately may improve quality of life and prevent sudden death. Delayed conduction on surface electrocardiogram was found to be potentially helpful for identifying patients at risk for sudden death or pacemaker implantation.4 Similarly with the other MD where cardiac involvement is not uncommon, early detection of underlying asymptomatic cardiac involvement is necessary to maintain cardiac function and prevent sudden death.


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<thead>
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<tbody>
<tr>
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<td>• Quality improvement&lt;br&gt;• Accountability</td>
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<td>Level of Measurement</td>
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<tr>
<td>Care setting</td>
<td>• Inpatient Services&lt;br&gt;• Outpatient visits&lt;br&gt;• Nursing Homes&lt;br&gt;• Rehabilitation Services&lt;br&gt;• Home Care Services</td>
</tr>
<tr>
<td>Data source</td>
<td>• Electronic health record (EHR) data&lt;br&gt;• Administrative Data/Claims (inpatient or outpatient claims)&lt;br&gt;• Administrative Data/Claims Expanded (multiple-source)&lt;br&gt;• Paper medical record</td>
</tr>
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**Technical Specifications: Administrative/Claims Data (Under Development)**

Administrative claims data collection requires users to identify the eligible population (denominator) and numerator using codes recorded on claims or billing forms (electronic or paper). Users report a rate based on all patients in a given practice for whom data are available and who meet the eligible population/denominator criteria.

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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)
**Measure Description**

All visits for patients with a diagnosis of a muscular dystrophy where the patient had a scoliosis evaluation* ordered.

**Measure Components**

| Numerator Statement | Patients who had a scoliosis evaluation ordered.*  
*Scoliosis evaluation: clinical evaluation, x-rays ordered, referral for orthopedic consultation or to a qualified clinician, etc. |
|---------------------|------------------------------------------------|
| Denominator Statement | All visits for patients with a diagnosis of a muscular dystrophy.  
Denominator Exceptions:  
- Medical reason for not ordering a scoliosis evaluation (eg patient cannot tolerate evaluation, MD phenotype not associated with scoliosis, etc.)  
- Patient reason for not ordering a scoliosis evaluation (eg patient or family caregiver declines evaluation)  
- System reason for not ordering a scoliosis evaluation (eg patient has no insurance coverage for x-rays or referral for consultation evaluation) |
| Supporting Guideline & Other References |  
- I1. Clinicians should monitor muscular dystrophy patients for the development of spinal deformities to prevent resultant complications and preserve function. (Level B)¹  
- I2. Clinicians should refer muscular dystrophy patients with musculoskeletal deformities of the spine to an orthopedic spine surgeon for monitoring and surgical intervention if deemed necessary to maintain normal posture, assist mobility, maintain cardiopulmonary function, and optimize quality of life. (Level B)¹  
- 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]²  
- Children with NMW who require surgery (including scoliosis surgery) should be assessed by a multidisciplinary team prior to any intervention. [GPP]²  
- The effect of wearing a spinal brace on respiratory function should be assessed and weighed against the limited evidence of benefit in terms of affecting final scoliosis severity. [D]²  
- The primary consideration when planning surgery for children with scoliosis associated with NMW should be comfort and quality of life. [GPP]²  
- Spinal care should involve an experienced spinal surgeon, and comprises scoliosis monitoring, support of spinal/pelvic symmetry and spinal extension by the wheelchair seating system, and (in patients using glucocorticoids, in particular) monitoring for painful vertebral body fractures. (Consensus)³  
- Monitoring for scoliosis should be by clinical observation through the ambulatory phase, with spinal radiography warranted only if scoliosis is observed. In the non-ambulatory phase, clinical assessment for scoliosis is essential at each visit. Spinal radiography is indicated as a baseline assessment for all patients around the time that wheelchair dependency begins with a sitting anteroposterior full-spine radiograph and lateral projection film. An anteroposterior spinal radiograph is warranted |
annually for curves of less than 15° to 20° and every 6 months for curves of more than 20°, irrespective of glucocorticoid treatment, up to skeletal maturity. (Consensus)³

- It is important to find out whether a child with DMD belongs to the small minority that does not develop a severe scoliosis. For this purpose, the respiratory functions should be monitored in children with DMD, since the vital capacity is a possible indicator of the progression of scoliosis. (Level 2)⁴


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Rationale for the Measure
There is a risk of evolving musculoskeletal spine deformities, such as scoliosis, kyphosis, or rigid spine syndrome, in various dystrophies. These musculoskeletal deformities can result in discomfort and functional impairment, interfering with gait, activities of daily living, and pulmonary function. The proper management of musculoskeletal spine deformities is important in order to reduce discomfort, preserve mobility or ability to sit in a wheelchair, and reduce pulmonary complications.¹

Gap in Care
There is limited data on a gap in care for scoliosis evaluation with a marked absence of randomized controlled trials on the evaluation or treatment of scoliosis. However, severe scoliosis causes discomfort, pain and compromises respiratory function. Surgery is the primary treatment for scoliosis but there are uncertainties as to the necessity and timing of the surgery.

Opportunity for Improvement
The Dutch Guideline on the Treatment of Scoliosis in DMD focused on recommendations for professionals managing the care of patients with scoliosis due to neuromuscular disease, DMD or SMA2.² The guideline supports multidisciplinary approach and encourages collaboration between the different specialties involved.


Do Not Cite. For Public Comment Period

- Accountability

**Type of measure**
- Process

**Level of Measurement**
- Individual practitioner

**Care setting**
- Outpatient visits
- Nursing Home
- Home Services
- Rehabilitation Services

**Data source**
- Electronic health record (EHR) data
- Administrative Data/Claims (inpatient or outpatient claims)
- Administrative Data/Claims Expanded (multiple-source)
- Paper medical record

**Technical Specifications: Administrative/Claims Data (Under Development)**
Administrative claims data collection requires users to identify the eligible population (denominator) and numerator using codes recorded on claims or billing forms (electronic or paper). Users report a rate based on all patients in a given practice for whom data are available and who meet the eligible population/denominator criteria.

The specifications listed below are those needed for performance calculation. Additional CPT II codes may be required depending on how measures are implemented. (Reporting vs. Performance)

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**AND**
CPT E/M Service Code:
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);
99307, 99308, 99309, 99310 (Subsequent Nursing Facility Care);
99319 (Other Nursing Facility Services);
DRAFT MEASURE #6: Patient Referred for Physical, Occupational or Speech/Swallowing Therapy

**MUSCULAR DYSTROPHY**

**Measure Description**

All visits for patients diagnosed with a muscular dystrophy where the patient was referred for physical, occupational or speech/swallowing therapy.

**Measure Components**

<table>
<thead>
<tr>
<th>Numerator Statement</th>
<th>Patient visits where the patient was referred for physical, occupational, or speech/swallowing therapy.</th>
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<tbody>
<tr>
<td>Denominator Statement</td>
<td>All visits for patients diagnosed with a muscular dystrophy.</td>
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</table>
| Denominator Exceptions | Exceptions:
  - Medical exception for not referring for physical, occupational, or speech/swallowing therapy. (eg patient does not need therapy based upon MD phenotype; patient already currently being seen by PT, OT or speech/swallowing specialist)
  - Patient exception for not referring for physical, occupational, or speech/swallowing therapy. (eg patient or family caregiver declines)
  - System exception for not referring for physical, occupational, or speech/swallowing therapy. (eg patient does not have insurance to cover therapy, patient cannot travel to the therapy facility) |
| Supporting Guideline & Other References | • L5. In patients with muscular dystrophy, clinicians should prescribe physical and occupational therapy, as well as bracing and assistive devices that are adapted specifically to the patient's deficiencies and contractures to preserve mobility and function and prevent contractures (Level B)
• L3. Clinicians should recommend that patients with muscular dystrophy have periodic assessments by a physical and occupational therapist for symptomatic and preventative screening. (Level B)
• F1. Clinicians should refer muscular dystrophy patients with dysphagia, frequent aspiration or weight loss for speech therapy and/or gastroenterology evaluation to assess and manage swallowing function, aspiration risk, teach patients techniques for safe and effective swallowing (“chin tuck” maneuver, altered food consistencies, etc.) and to consider placement of gastrostomy/jejunostomy tube for nutritional support. (Level B)
• L1. Clinicians should refer patients with muscular dystrophy to a clinic that has access to multiple specialties (e.g., Physical Therapy, Occupational Therapy, Respiratory Therapy, Speech and swallowing Therapy, cardiology, pulmonology, orthopedics and genetics ) designed specifically to care for patients with muscular dystrophy and other neuromuscular disorders in order to provide efficient and effective long-term care. (Level B)
• H1. Physicians should consider referrals to allied health professionals, including physical, occupational, and speech therapists; seating and mobility specialists; rehabilitation specialists; and orthopaedic surgeons, to help maximize function and potentially slow the progression of musculoskeletal complications in children with CMD (Level TBD).
• D3. Clinicians should encourage patients with FSHD to engage in low intensity aerobic exercise. Clinician can use the practical physical activities guidelines for individuals with disabilities provided by the department of Health and Human Services (http://www.health.gov/paguidelines/guidelines/chapter7.aspx) to counsel patients about aerobic exercise. |
• D4. For patients interested in strength training, clinicians should refer patients to physical therapists to establish a safe exercise program using appropriate low/medium weights/resistance that takes into consideration the patient’s physical limitations.3

• Children with neuromuscular disease with a history of swallowing difficulties should have a feeding assessment by a speech and language therapist including a video fluoroscopy swallow assessment if the swallow is thought to be unsafe. [O]4


Rationale for the Measure
Patients with muscular dystrophy may have difficulty receiving adequate oral intake due to dysphagia and/or inability to feed themselves due to excessive arm weakness. Maintaining adequate nutrition and body weight is important for optimizing strength, function, and quality of life. When oral intake is inadequate, other means of maintaining intake, such as gastrostomy or jejunostomy feeding tubes, may be needed to maintain optimal nutrition. There is evidence from related conditions (amyotrophic lateral sclerosis [ALS]) that maintenance of nutrition and body weight prolongs survival.1

The principles of the long-term management of patients with LGMD must emphasize maintaining mobility and functional independence for as long as possible, with a focus on maximizing quality of life. The prevention and management of comorbidities, both expected and acquired, is a major part of such management. This would include joint contractures, scoliosis, osteoporosis, dysphagia, and restrictive lung disease (expected), as well as obesity, metabolic syndrome, and stress fractures (acquired).1

Despite inadequate research in this area, the available evidence suggests that this population would benefit from both strengthening and aerobic fitness training programs. Due to the muscle degeneration in muscular dystrophy, there may be some risk of exercise-induced muscle damage and subsequent overwork weakness following supramaximal, high-intensity exercise. Overwork weakness is defined as a prolonged decrease in absolute muscle strength and endurance following strenuous or excessive exercise. It is often accompanied by extreme delayed onset muscle soreness, peaking 1-5 days postexercise, and possibly inducing myoglobinuria. Clinicians need to be prudent in their recommendations, encouraging alternating periods of physical activity and scheduled rest. Clinicians should also be aware that true overwork weakness has not been demonstrated in any trial of exercise done in this
population to date. All forms of physical exercise should therefore be prescribed cautiously, using a common sense approach. There have been several randomized or quasi-randomized controlled trials comparing strength training programs, aerobic exercise programs, or both to non-training controls in patients with a variety of neuromuscular disorders. On the basis of this literature, both strength training and aerobic exercise programs appear to be safe, without any notable deleterious effects.\(^1\)

**Gap in care**

Physical therapy should be started as early as possible. From the time of diagnosis, preventive therapy is an essential part of daily management. Referral to physical or occupational therapy is prompted by the diagnosis of muscular dystrophy, appearance of contractures, loss of motor function, decreased mobility, altered gait, abnormal positioning, muscle weakness, pain, scoliosis, problems with transfers, joint deformity, and loss of activities of daily living.\(^2\) Publications have emphasized the importance of rehabilitation in the management of pain.

Most medical centers do have physical, occupational and speech/swallowing therapists. The percentage of patients that do not get the needed physical therapy, occupational therapy or speech/swallowing therapy is unclear as studies have not been conducted to look at this specific gap in care. However, anticipatory guidance is needed by all three services to avoid functional deterioration and malnutrition.

**Opportunity for Improvement**

Physical therapy should focus on the maintenance of function and mobility, prevention or treatment of joint contractures and spine deformities, training of patients to carry out activities that are safe. PT can also recommend transfer aids and adaptive equipment to ensure the highest degree of independence and safety.

Occupational therapy (OT) should focus on encouraging patient to perform activities of daily living to the best ability. OT can also encourage patient to engage in activities such as singing or playing wind instruments, which may improve pulmonary function. OT also teaches the patient to maintain adequate seating position and wheelchair support. Such early and adequate posturing of feet and neck can effectively prevent foot deformities and hyperextension of the neck.

Speech and language pathologists assess MD patients for any swallowing difficulties, nutrition status and perform swallowing surveillance, deciding texture of food so to avoid aspiration. Identification and assessment of feeding difficulties are essential for optimal care of patients with a muscular dystrophy. Speech therapy intervention should focus on compensatory communication strategies, as necessary.

This quality measure has the potential with appropriate referral to for these types of therapy to improve quality of life and may length of life in people who have a muscular dystrophy.


Do Not Cite. For Public Comment Period

**Measure Designation**

| Measure purpose                  | • Quality improvement  
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| **Care setting**                 | • Inpatient Consultations  
|                                  | • Outpatient visits  
|                                  | • Nursing Homes   
|                                  | • Home Services   
|                                  | • Rehabilitation Services  |

**Technical Specifications: Administrative/Claims Data (Under Development)**

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99304, 99305, 99306 (Initial Nursing Facility Care)
99307, 99308, 99309, 99310 (Subsequent Nursing Facility Care)
99319 (Other Nursing Facility Services)
99324, 99325, 99326, 99327, 99328 (Domiciliary, Rest Home, or Custodial Care Services-New Patient)
99334, 99335, 99336, 99337 (Domiciliary, Rest Home, or Custodial Care Services-Established Patient)
99339, 99340 (Domiciliary, Rest Home, or Home Care Plan Oversight Services)
99341, 99342, 99343, 99344, 99345 (Home Services-New Patient)
99347, 99348, 99349, 99350 (Home Services-Established Patient).
**Measure Description**

All visits for patients diagnosed with muscular dystrophy where the patient’s nutritional status or growth trajectories were monitored.

**Measure Components**

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<th>Patient visits where the patient’s nutritional status or growth trajectories were monitored*.</th>
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<td>*Monitored defined as: referral for a nutrition or dietetic consultation, monitor weight, height (linear height in ambulatory patients and arm span/segmental length in non-ambulatory patients), muscle mass, BMI, growth charts, etc.</td>
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<td>• Medical reason for not monitoring for nutrition or growth trajectory problems or referring for these purposes (eg patient is already being following by a nutritionist or other qualified specialist for these issues)</td>
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<td>• Patient reason for not monitoring for nutrition or growth trajectory problems or referring for these purposes (eg patient or family caregiver declines)</td>
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<td></td>
<td>• System reason for not monitoring for nutrition or growth trajectory problems or referring for these purposes (eg patient is unable to travel)</td>
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| Supporting Guideline & Other References | • D2. The physician should refer the child with CMD to a pulmonary or aerodigestive care team that is experienced in managing the interface between oro-pharyngeal function, gastric reflux and dysmotility, nutrition and respiratory systems and can provide anticipatory guidance around trajectory, assessment modalities, complications and potential interventions (Level TBD).¹ |
|                                         | • E1. Neuromuscular specialists should coordinate with primary care providers to follow nutrition and growth trajectories (Level B).¹ |
|                                         | • In DMD, ensure adequate intake of micronutrients as per dietary reference values (Grade D)² |
|                                         | • In DMD, various tools can be used to measure body composition with DXA and MRI being accurate, appropriate and noninvasive measurement instruments (Grade C)² |
|                                         | • In DMD, measure height and weight every six months and plot on standard growth charts (Grade D)² |
|                                         | • In DMD, upper arm length, tibial length, or knee height can be measured in the advanced stage of disease (Grade D)² |
|                                         | • A problem-orientated approach to nutrition should aim to minimize risk of aspiration, optimize nutritional status, promote comfort and balance the positive social consequences of continued oral feeding. [O³] |
|                                         | • When adequate nutrition cannot be safely accomplished with oral feedings, gastrostomy tube placement and enteral feedings under the guidance of a nutritionist is strongly recommended. ⁴ |
|                                         | • Percentage ideal body weight and body mass index must be assessed regularly and counseling provided as necessary.⁴ |
|                                         | • A nutritionist should evaluate patients with DMD as part of their regular follow-up care.⁴ |
Rationale for the Measure

Delayed growth, short stature, muscle wasting and increased fat mass are characteristics of DMD and impact on nutritional status and energy requirements. The early introduction of steroids has altered the natural history of the disease, but can exacerbate weight gain in a population already susceptible to obesity. Prior tocommencing steroids, anticipatory guidance for weight management should be provided. Malnutrition is a feature of end stage disease requiring a multidisciplinary approach, such as texture modification and supplemental feeding. As a result of corticosteroid treatment, vitamin D and calcium should be supplemented.\textsuperscript{1}

Patients with muscular dystrophy may have difficulty receiving adequate oral intake due to dysphagia and/or inability to feed themselves due to excessive arm weakness. Maintaining adequate nutrition and body weight is important for optimizing strength, function, and quality of life. When oral intake is inadequate, other means of maintaining intake, such as gastrostomy or jejunostomy feeding tubes, may be needed to maintain optimal nutrition. There is evidence from related conditions (amyotrophic lateral sclerosis [ALS]) that maintenance of nutrition and body weight prolongs survival.\textsuperscript{2}

Gap in Care

One of the problems in monitoring nutrition and growth is the absence of appropriate growth charts and data on energy and nutritional requirements in this population. The most encountered problem is under nutrition and poor weight gain.\textsuperscript{3,4} Overweight also has to be considered, particularly in the adult population because of the limited mobility of these patients.\textsuperscript{3,4} Growth should be screened by regular weight measurements, completed by height or a surrogate height measurement (arm span or ulnar length).\textsuperscript{3} Anticipatory guidance and prevention of undernutrition/malnutrition and being overweight/obese should be goals from diagnosis throughout life.\textsuperscript{4}

Opportunity for Improvement

Patients with congenital muscular dystrophy often have a growth curve below what is expected for age. This is acceptable if the child is in good health and has no signs of fatigue, recurrent infections, or cardiac and respiratory dysfunction. This underscores the need for regular assessment including detailed history taking for feeding issues and full examination. Despite the lack of appropriate growth charts, stagnated growth is a concern in a growing child, necessitating repeated measurements that can require nutritional interventions.\textsuperscript{3}

\textsuperscript{1} Davidson ZE, Truby H. A review of nutrition in Duchenne muscular dystrophy. \textit{J Hum Nutr Diet} 2009. 22(5):383-393
\textsuperscript{2} Davidson Z, Truby H.; A Review of Nutrition in Duchenne Muscular Dystrophy; \textit{J of Hum Nutr Diet} 2009. 22(5):383-393
\textsuperscript{4} American Thoracic Society. Respiratory Care of the Patient with Duchenne Muscular Dystrophy. \textit{Am J Respir Crit Care Med} 2004; (170):456-465. This is a consensus document; not a systematic review or a practice guideline.
Measure Designation

| Measure purpose         | • Quality improvement  
|                        | • Accountability      |
| Type of measure        | • Process             |
| Level of Measurement   | • Individual practitioner  |
| Care setting           | • Outpatient visits  
|                        | • Nursing homes  
|                        | • Rehabilitation Services  
|                        | • Home Care Services     |
| Data source            | • Electronic health record (EHR) data  
|                        | • Administrative Data/Claims (inpatient or outpatient claims)  
|                        | • Administrative Data/Claims Expanded (multiple-source)  
|                        | • Paper medical record

Technical Specifications: Administrative/Claims Data (Under Development)

Administrative claims data collection requires users to identify the eligible population (denominator) and numerator using codes recorded on claims or billing forms (electronic or paper). Users report a rate based on all patients in a given practice for whom data are available and who meet the eligible population/denominator criteria.

The specifications listed below are those needed for performance calculation.

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<td>359.22 Myotonia congenita</td>
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<tr>
<td>359.89 Other myopathies</td>
<td>G72.89 Other specified myopathies</td>
</tr>
<tr>
<td>------------------------</td>
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</tr>
<tr>
<td>359.9 Myopathy, unspecified</td>
<td>G72.9 Myopathy, unspecified</td>
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</tbody>
</table>

AND

CPT E/M Service Code:
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);
99307, 99308, 99309, 99310 (Subsequent Nursing Facility Care);
97001, 97002, 97003, 97004 (PT/OT evaluation)
99341, 99342, 99343, 99344, 99345 (Home Services-New Patient);
99347, 99348, 99349, 99350 (Home Services-Established Patient).
DRAFT MEASURE #8: Patient Queried about Pain and Pain Interference with Function
MUSCULAR DYSTROPHY

Measure Description
All visits for patients diagnosed with a muscular dystrophy where the patient was queried about pain and pain interference with function using a validated and reliable instrument*.

Measure Components

| Numerator Statement | Patient visits where the patient was queried about pain and pain interference with function using a validated and reliable instrument*.

*Note: Pain can be assessed using one of a number of available valid and reliable instruments available from medical literature. Examples, include, but are not limited to:
- Numeric Rating Scale for Pain
- Faces Pain Scale
- Graded Chronic Pain Scale
- Visual Analogue Scale
- McGill Pain Questionnaire
- Short-Form McGill Pain Questionnaire


Denominator Statement | All visits for patients diagnosed with a muscular dystrophy.

Denominator Exceptions
- Medical reason for not querying about pain and pain interference with function (eg patient is cognitively impaired and unable communicate)
- Patient reason for not querying about pain and pain interference with function (eg patient declines to respond to questions)

Supporting Guideline & Other References
- Routine pain evaluation should be part of standard clinical assessment in all children and young people with neuromuscular disorders. [Level D]¹
- C6. Treating physicians should routinely inquire about pain in patients with FSHD.²

Rationale for the Measure
Between 68-82% of patients with muscular dystrophies live in pain.\(^1\) Pain is a common feature of some MDs, notably myotonic dystrophy and FSHD, but also many of the LGMDs. Pain interferes with physical and psychological functioning in these patients. Lower extremity pain intuitively affects ambulation.\(^2,3\) Pain and fatigue are independent predictors of lower physical functioning and greater depression.\(^4\) Thus identification and treatment of pain is important to improve the care of patients with MD.

Gap in Care:
Pain in the back and the legs is most commonly reported. Most patients do not receive optimal and effective treatments.\(^3\) One paper reported that pain is multifactorial and can be a significant and under recognized problem in congenital muscular dystrophy.\(^5\) Effective management begins with a comprehensive assessment of acute and chronic pain to determine the presence, frequency, and duration of painful episodes and to identify alleviating or exacerbating factors.\(^6,7\)

Opportunity for Improvement:
A multitude of treatment modalities are available to control or relieve the pain using non-pharmacological, pharmacological, and interventional approaches in this patient group. Access to these treatments could improve the quality of life. Adequate assessment of pain using validated and easy-to-use tools to measure pain is a key step to bridge this gap. The Numeric Rating Scale for Pain and Faces Pain Scale are such tools and the compliance with measurement task is high.


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<table>
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<th>Measure Designation</th>
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</table>
| Measure purpose     | • Quality improvement  
|                     | • Accountability  |
| Type of measure     | • Process  |
| Level of            | • Individual practitioner  |
Measurement

<table>
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<tbody>
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Technical Specifications: Administrative/Claims Data (Under Development)

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CPT E/M Service Code:
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**Measure Description**

All patients with a diagnosis of a muscular dystrophy (or caregivers) who were counseled about advanced health care decision making, palliative care or end-of-life issues at least once annually.

**Measure Components**

<table>
<thead>
<tr>
<th>Numerator Statement</th>
<th>Patients (or caregivers) who were counseled about advanced health care decision-making, palliative care, or end-of-life issues* at least once annually.</th>
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<td>*Advanced health care decision making, palliative care and end-of-life issues may include: emotional, spiritual, developmental, or physical dimensions.</td>
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<th>All patients with a diagnosis of a muscular dystrophy.</th>
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<tr>
<th>Denominator Exceptions</th>
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<tbody>
<tr>
<td></td>
<td>• Medical exception for not counseling about advanced health care decision making, palliative care or end of life issues (e.g. patient is unable to communicate and caregiver is not available)</td>
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</table>

| Supporting Guideline & Other References | • L4. While respecting and protecting patient autonomy, the clinician should proactively anticipate and facilitate the patient and family in making the necessary decisions required as the disease progresses, including loss of mobility, need for assistance with activities of daily living, medical complications and end-of-life care. (Level B)¹ |
|                                        | • Families need access to skilled experts for multidimensional coordinated palliative care support, providing regular review of their needs at various stages in their condition. [D]² |
|                                        | • Generic palliative care skills should be cascaded to other professionals providing neuromuscular services. [O]² |
|                                        | • Written plans for the management of acute exacerbations, which include details of effective airway clearance methods and ventilator settings when appropriate, and contact details of relevant healthcare professionals are recommended. [O]² |
|                                        | • Assisting patients, parents and carers to make informed choices that are consistent with their own values and preferences requires physicians to engage patients and their parents and carers in a process of mutual participation in decision-making with full disclosure of all information in a sensitive and timely fashion (D)² |
|                                        | • Advance care planning should be an integral part of the active management of children and young people with neuromuscular disorders. Advance care plans can be used as a vehicle for information exchange and considered decision-making, [D]² |
|                                        | • Patients and families need to have ownership of the advance care plan and be educated as to its uses. [D]² |
|                                        | • Advance care plans should be reviewed by the multidisciplinary team on a regular basis. [GPP]² |
|                                        | • Generic palliative care skills should be cascaded to other professionals providing neuromuscular services. [GPP]² |
|                                        | • End of life decision-making requires the provision of adequate information to the patient and family.³ |
|                                        | • Physicians must actively work collaboratively with the patient, family members |
and other health professionals involved in the health care decision-making process while at all times maintaining respect for patient autonomy, dignity and confidentiality. (Consensus)\(^4\)

- It is important to proactively counsel capable patients and establish clear advanced directives (regarding issues such as crisis management and end-of-life care) in a timely manner, ensuring that patients fully understand and appreciate the reasonably foreseeable outcomes of their decisions. Physicians must work with patients to help prioritize their values, interests and preferences. (Consensus)\(^4\)

- When considering the most appropriate location for ongoing ventilation issues relating to safety and the patient’s values, beliefs and preferences must be the primary considerations for making such decisions providing optimal independence, respect for patient autonomy and increased quality of life. (Consensus)\(^4\)

- One must recognize one’s own biases and endeavor to participate in a collaborative and fair decision-making process that primarily addresses, reflects and respects the values and wishes of the patient. (Consensus)\(^4\)


3 American Thoracic Society. Respiratory Care of the Patient with Duchenne Muscular Dystrophy. *Am J Respir Crit Care Med* 2004; (170):456-465  This is a consensus document; not a systematic review or a practice guideline.


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**Rationale for the Measure**

An important aspect of ongoing management includes proactively preparing patients with muscular dystrophy and their families for the long-term consequences of muscular dystrophies and engaging in discussions regarding end-of-life care. This helps patients come to terms with their condition and prepare for the expected complications of their form of muscular dystrophy and avoids the need for hasty decisions made in the throes of a medical crisis.\(^1\) Palliative care is useful to alleviate the suffering of these patients.\(^2\)

**Gap in Care**

Families of children with life-limiting conditions who are on long-term assisted ventilation need to undertake end-of-life advance care planning (ACP) in order to align their goals and values with the inevitability of their child's condition and the risks it entails.\(^3\) By offering anticipatory guidance and encouraging contemplation of patients’ goals both in times of stability and during worsening illness, health care providers can better engage patients’ families in ACP.\(^3\) As the child's condition progresses, the emphasis can be recalibrated. How families respond to such encouragement can also serve as a gauge of their willingness to pursue ACP.

In one study of palliative care services for male patients with DMD (n=34) 85% of families had never heard the term palliative care. Only attendant care and skilled nursing services showed much usage, with 44% and 50% indicating receipt of these
services, respectively. Receipt of other services was reported less frequently; pastor care (27%), respite care (18%), pain management (12%), and hospice care (6%). Only 8 respondents (25%) reported having any type of directive document in place.  

**Opportunity for Improvement**

Health care providers should educate patients and families that palliative care is complementary to care with curative intent and that incorporates palliative care principles during ongoing therapies will improve support systems during illness.

Comprehensive care for congenital muscular dystrophies should encompass the entire life span, and a clear distinction should be made between “life-limiting” diagnosis and a “life-threatening” episode, considering that the trajectory of life toward death will be highly variable and certainly individual. Incorporating palliative care from diagnosis can benefit the patient, family, and medical team as they anticipate and make decisions regarding interventions that affect both the duration and quality of these individuals’ lives.  

Results of the congenital muscular dystrophy Family Standard of Care survey indicate that families prefer to be made aware of potential outcomes of the congenital muscular dystrophy diagnosis across medical disciplines, not just with end-of-life discussions. This information can serve as a platform to discuss choices that are available for common life-threatening complications before they occur.

It is the responsibility of the providers to initiate end of life discussions and to provide families with information regarding options for care. This should happen before the occurrence of a major life-threatening event, allowing families time to clearly explore options and gather information before a decision must be made. The goal is to partner with families to present them with information in a developmentally appropriate and culturally sensitive manner while elucidating that their choices may change at any time. A written plan should be developed that clearly states the parents’ and child’s wishes for both emergency situations and slower illness deterioration, as this will allow families to feel more in control during these times.  


### Measure Designation

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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);
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- 99347, 99348, 99349, 99350 (Home Services-Established Patient).
- 97001, 97002, 97003, 97004 (PT/OT evaluation)
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