American Academy of Neurology

Muscular Dystrophy

Quality Measurement Set

Final

Approved by the Muscular Dystrophy Quality Measurement Development Work Group on February 25, 2014, by the AAN Quality and Safety Subcommittee on March 13, 2014; by the AAN Practice Committee on April 8, 2014; and by the AANI Board of Directors on May 28, 2014.

Clinician Performance Measures (Measures) and related data specifications developed by the American Academy of Neurology (AAN) are intended to facilitate quality improvement activities by clinicians.

These measures are intended to assist clinicians in enhancing quality of care. Measures are designed for use by any clinician 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.

Measures are subject to review and may be revised or rescinded at any time by the AAN. The measures may not be altered without prior written approval from the AAN. The measures, while copyrighted, can be reproduced and distributed, without modification, for noncommercial purposes (e.g., use by health care providers in connection with their practices). Commercial use is defined as the sale, license, or distribution of the measures for commercial gain, or incorporation of the measures into a product or service that is sold, licensed, or distributed for commercial gain. Commercial uses of the measures require a license agreement between the user and the AAN. Neither the AAN nor its members shall be responsible for any use of the measures.

THESE MEASURES AND SPECIFICATIONS ARE PROVIDED "AS IS" WITHOUT WARRANTY OF ANY KIND.

This publication was supported by the Cooperative Agreement Number (Grant Number: #5U38DD000723-03) from the Centers for Disease Control and Prevention. Its contents are solely the responsibility of the authors and do not necessarily represent the official views of the Centers for Disease Control and Prevention.

Limited proprietary coding is contained in the measure specifications for convenience. Users of the proprietary coding sets should obtain all necessary licenses from the owners of these code sets. The AAN and its members disclaim all liability for use or accuracy of any Current Procedural Terminology (CPT®) or other coding contained in the specifications.

American Academy of Neurology

Muscular Dystrophy Measure Development Work Group

Co-Chairs
John D. England, MD, FAAN
Pushpa Narayanaswami, MBBS, MD, FAAN

Physician or Clinician Society/ Associations

American Academy of Neurology
Paula Clemens, MD
William S. David, MD
Gary Gronseth, MD
Carla Grosmann, MD
Michelle Mellion, MD
Jonathan Strober, MD

American Association of Neuromuscular and Electrodiagnostic Medicine
Benn Smith, MD

American Occupational Therapy Association
Roxanna Bendixen, PhD, OTR

American Academy of Orthopedic Surgeons
Frederic Shapiro, MD

American Academy of Pain Medicine
Jiangu Cheng, MD, PhD

American Academy of Physical Medicine and Rehabilitation
Robert Rinaldi, MD

American College of Cardiology
Christopher F. Spurney, MD

American Physical Therapy Association
Shreedevi Pandya, PT, DPT, MS

American Thoracic Society
Jonathan Finder, MD

Child Neurology Society
Ryan Coates, MD

Patient Advocacy Organizations

Muscular Dystrophy Association
Jodi Wolff, MSSW

Cure CMD
Meganne Leach, MSN, APRN, PNP-BC

Insurers
Blue Cross Blue Shield
Frank Lefevre, MD

Work Group Facilitators (Non-Voting):
Richard Dubinsky, MD, MS
David Wang, DO

American Academy of Neurology Staff:
Rebecca Swain-Eng, MS, CAE
Gina Gjorvad
Becky Schierman, MPH

DECLINED: American Academy of Family Physicians, American College of Physicians, American College of Emergency Physicians, American Osteopathic Association, Aetna, UnitedHealth Care, Humana, Palmetto GBA, Highmark, Cigna, Kaiser Permanente, Wellpoint, Myotonic Dystrophy Foundation, MN Health Action Group, Midwest Business Group on Health, Academy of Nutrition and Dietetics, National Hospice and Palliative Care Organization

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 (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 (DMD), congenital muscular dystrophy, facioscapulohumeral muscular dystrophy (FSHD) and limb-girdle muscular dystrophy (LGMD). 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

- To estimate the population-based prevalence of Duchenne/Becker muscular dystrophy (DBMD) and describe selected clinical outcomes, The Centers for Disease Control and Prevention (CDC) and investigators from the Muscular Dystrophy Surveillance Tracking and Research Network (MD STARnet) analyzed data for males born during 1983--2002 that were reported to the MD STARnet from four participating states. Since 2004, MD STARnet has conducted named population-based surveillance of DBMD in four states (Arizona, Colorado, Iowa, and 12 counties* in western New York) for males born on or after January 1, 1982. Overall DBMD prevalence rates per 10,000 males aged 5--24 years ranged from 1.3 (Arizona) to 1.8 (western New York). Age-and state-specific prevalences per 10,000 males ranged from 0.9 (Iowa) to 1.9 (western New York) for males aged 5--9 years; 1.4 (Colorado) to 2.5 (Iowa) for males aged 10--14 years; 1.6 (Arizona) to 2.5 (Colorado) for males aged 15--19 years; and 0.8 (Arizona) to 1.1 (western New York) for males aged 20--24 years. For the 349 males with DBMD at the beginning of 2007, the age-specific percentages for those who used wheelchairs were 29% at age 5--9 years and >90% at age ≥15 years.[1]
- Birth prevalence of DMD has been estimated at 1 in 3,500 (2.9 per 10,000) male births and Becker muscular dystrophy (BMD) at 1 in 18,518 (0.5 per 10,000) male births. Emery AE. Population frequencies of inherited neuromuscular diseases---a world survey.[2].
- The most common form, DMD, affects 1 in every 3,500 to 6,000 male births each year in the United States. DMD accounts for approximately 50 percent of all cases.[3]
- Becker Muscular Dystrophy (BMD)-affects males. Becker muscular dystrophy 1/18,000–1/31,000[4]
- Congenital Muscular Dystrophy (CMD)-affects both males and females. 2.5/100,000[4]
- Distal Muscular Dystrophy (DD): Most inherited in an autosomal dominant pattern but some follow a recessive pattern of inheritance. 1/10,000[4]
- Emery-Dreifuss Muscular Dystrophy: Primarily affects males. 1/100,000[4]
- Facioscapulohumeral Muscular Dystrophy (FSHD)-3rd most common form of MD 1/20,000.[4]
- Myotonic dystrophy. Common form of MD. 1/8,000[4]
- Oculopharyngeal Muscular Dystrophy (OPMD)-late onset. It's autosomal dominant or autosomal recessive. 1/1000 in French Canadians and highest in Uzbekistan Burkhara Jews with 1:600.[4,5]

1. Romitti P, Puzhankara S, Mathews K, et al. Prevalence of Duchenne/Becker Muscular Dystrophy Among Males Aged 5-24 Years – Fou States, 2007. MMWR 2009;58(40):1119-1122. Available at: http://www.cdc.gov/mmwr/preview/mmwrhtml/mm5840a1.htm Accessed on February 15, 2014.
2. Emery AE. Population frequencies of inherited neuromuscular diseases---a world survey. Neuromuscul Disord 1991; 1:19—29.
3. Centers for Disease Control and Prevention, National Center on Birth Defects and Developmental Delays, July 17, 2013.
4. Blumen SC, Nisipeanu P, Sadeh M, *et al.* Epidemiology and inheritance of oculopharyngeal muscular dystrophy in Israel. *Neuromuscul. Disord.* 1997 Oct 7;7(Suppl 1): S38–40.
5. Banwell BL. 223 Muscular Dystrophies. http://www.macpeds.com/documents/Neuromuscular-MuscularDystrophies-BanwellChapter.pdf Accessed on February 15, 2014.

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 trunk. The heart and respiratory muscles can also be affected.
- Some types of MD shorten 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):
    - BMD-survival is usually into old age.
    - CMD-shortened life span.
    - DMD-life span ranges from 15-to-51 years. Muscle degeneration may be mild or severe.
    - Distal MD-onset at ages 20-to-60 years old. Progress is slow and not life threatening.
    - Emery-Dreifuss muscular dystrophy-present in childhood and early teenage years with contractures. At risk for stroke and sudden death from cardiac complications.
    - FSHD-affects muscles of the factors, shoulders, and upper arms with progressive weakness. Some affected individuals become severely disabled.
    - Myotonic dystrophy-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.
    - OPMD-onset at ages 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 (QOL) can be rated in terms of physical disability, pulmonary function, mobility, independence, etc.
- A comparison of Health Related Quality of Life (HRQoL) of patients with MD using validated instruments in different age groups indicated that having MD negatively influences the HRQoL on several domains (e.g., physical symptoms, motor functioning, autonomy, cognitive functioning, social functioning, positive emotions and negative emotions).[1] .)
- 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.[2]
- 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.[3]

1. Grootenhuis MA, De Boone, JD, Van Der Kooi A. Living with muscular dystrophy: health related quality of life consequences for adults and children. Health and Quality of Life Outcomes 2007 June;5:31-38. Available at http://link.springer.com/article/10.1186%2F1477-7525-5-31#page-2 Accessed on February 15, 2014.
2. Uzark K, King E, Cripe L, et al. Health-Related Quality of Life in Children and Adolescents with Duchenne Muscular Dystrophy. Pediatrics 2012;130(6):e1559-e1566. Available at: http://pediatrics.aappublications.org/content/130/6/e1559 Accessed on February 15, 2014.
3. Kohler M, Clarenbach CF, Böni L, et al. Quality of Life, Physical Disability, and Respiratory Impairment in Duchenne Muscular Dystrophy. American Journal of Respiratory and Critical Care Medicine 2005;172(8):1032-1036. Available at: http://www.atsjournals.org/doi/full/10.1164/rccm.200503-322OC Accessed on February 15, 2014.

Costs

- Medical costs are largely driven by outpatient care. Non-medical costs were driven by the necessity to move or adapt housing for the patient and paid caregiving. Annual per-patient costs for DMD range from$50,952 to $32,236 for Myotonic dystrophy. Population wide-national costs were $787 million (DMD) and $448 million (Myotonic dystrophy).[1]

- The yearly average cost in 2004 for medical care for privately insured individuals with any type of MD was $18,930; ranging from $13,464 at 5 through 9 years of age to $32,541 at 15 through 19 years of age.[2]
- In 2005, the financial cost of MD was $435 million. Of this:
    - $236.2 million (54.2%) was productivity lost due to lower employment, absenteeism and premature death of Australians with MD;
    - $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;
    - $42.4 million (9.7%) was the deadweight loss from transfers including welfare payments (Mainly Disability Support Pension and Career Payment) and taxation forgone;
    - $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;
    - $7.4 million (2.2%) was the direct health system expenditure.
- In per capita terms, this amounts to a financial cost of around $126,000 per person with MD per annum. Including the value of lost wellbeing, the cost is over $415,000 per person per annum.[3]

1. Larkindale J, Yang W, Hogan PF, et al. Cost of illness for neuromuscular disease in the U.S. Muscle Nerve 2014 Mar;49(3):431-438.
2. Centers for Disease Control and Prevention. Muscular Dystrophy Data and Statistics. Available at: http://www.cdc.gov/ncbddd/musculardystrophy/data.html Accessed on February 15, 2014.
3. Access Economics for the Muscular Dystrophy Association. The cost of Muscular Dystrophy. October 2007 Report. Available at: http://www.mda.org.au/media/accesslaunch/ExecutiveSummary5.pdf Accessed on February 15, 2014.

Gaps in Care and Opportunities for Improvement

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

Disparities

- MD occurs worldwide and affects all races.
- The age adjusted mortality rate for MD-associated deaths was lower in blacks (0.251, 95% CI: 0.205–0.297) than in whites (0.388, 95% CI: 0.368–0.409. Autosomal and X-linked MDs each accounted for approximately half of the deaths associated with MD. The mortality rate was higher for Whites (0.388 per 100,000 persons per year) than for Blacks (0.251 per 100,000 persons per year). Potential reasons for this difference include different prevalence rates, differential rates of diagnosis, and differences in reporting on death certificates. In addition to differences in mortality rates, there were also differences in the median age at death between Blacks and Whites. The median age at death was lower in Blacks than in Whites for both males and females. This could be due to (1) differences in mixtures of the types of MDs; (2) differences in ascertainment (in Blacks, only the more severe cases might be more likely than less severe cases to be recorded on death certificates); or (3) actual differences in age at death in individuals with comparable MDs. The increase over time in age at death for White males but not Black males might indicate that the differences reflect actual differences in age at death, perhaps due to inequities in access to health care or other factors. Cardiac complications were more commonly noted among MD-associated deaths in Blacks (38.9%) than Whites (28.6%). Another possible explanation for the lower age at death for Blacks than Whites is the increased frequency of cardiac complications reported.[1]

1. Kenneson A, Kolor K, Yang Q, et al. Trends and racial disparities in muscular dystrophy deaths in the United States, 1983-1998: an analysis of multiple cause mortality data. AM J Med Genet A. 2006 Nov 1;140(21):2289-2297. Available at: http://www.ncbi.nlm.nih.gov/pubmed/17022078 Accessed on February 15, 2014.

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 DMD, CMD, FSHD and LGMD. Therefore this measurement set is predominantly focused on these types of MD.

Evidence papers from the American Academy of Neurology, Mullender, et al. (2008), Bushby, et al. (2005, 2009, 2010), British Thoracic Society (2012), Wolfe, et al. (2012), American Thoracic Society (2004, 2009 Update Verified by Finder, et al.), American Academy of Pediatrics (2009 Affirmed), Andrews, et al. (2013), Scully, et al. (2013), Davidson, et al. (2009), McKim (2011), and other peer-reviewed publications in specific areas of MD treatment or management where rigorous guidelines did not exist were used as the evidence base for the measures in this measurement set.

Muscular Dystrophy Outcome Quality Measures

The workgroup thoroughly discussed many desired outcomes for the care of patients diagnosed with a MD. (See the list below under "Desired Outcomes for Patients with a Muscular Dystrophy".) The Work Group drafted two outcome measures that 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 voted down and dropped from the draft measurement set primarily because of the lack of strong guideline-based, high-level evidence recommendations or feasibility issues.

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, and anxiety
2. Independence
    a. Preserve or improve ambulation (focused on DMD)
        i. Measure timed walk
        ii. Capture age at time different ambulation aids are used (i.e., cane, walker, wheelchair, powered wheel chair)
    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 (Activities of Daily Living) - physical or occupational therapy, exercise programs, orthoses
        i. Measure participation in exercise programs
        ii. Functional independence measures
    d. Communication
        i. Prevent and/or reduce social isolation
    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 MD
        i. Utilization of muscle biopsy
        ii. Utilization of genetic tests 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 LGMD, FSHD)
        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 (e.g., sleep apnea, cataracts, diabetes in DM1, retinopathy, deafness in FSHD, learning disabilities or cognitive dysfunction in DMD and some other dystrophies such as dystroglycanopathies)
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
    a. 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
        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)

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 MD. 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.

AAN Muscular Dystrophy 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. Patient Referred for Physical, Occupational, or Speech/Swallowing Therapy
7. Nutrition Status or Growth Trajectories Monitored
8. Patient Queried about Pain and Pain Interference with Function
MD Planning and Patient Engagement
9. Patient 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 purse 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.

Measure	Safe	Effective		Patient-Centered	Timely	Efficient	Equitable
		Underuse	Overuse				
1. Patients with DMD Prescribed Appropriate Disease Modifying Pharmaceutical Therapy	X	X		X	X		
2. MD Multidisciplinary Care Plan Developed or Updated	X	X		X	X	X	X
3. Evaluation of Pulmonary Status Ordered	X	X		X	X		X
4. Evaluation of Cardiac Status Ordered	X	X		X	X		X
5. Scoliosis Evaluation Ordered.	X	X		X	X		X
6. Patient referred for Physical, Occupational, or Speech/Swallowing Therapy	X	X		X	X		
7. Nutrition Status or Growth Trajectories Monitored	X	X		X	X		
8. Patient queried about Pain and Pain Interference with Function	X	X		X	X		
9. Patient counseled about Advanced Health Care Decision Making, Palliative Care, or End of Life Issues	X	X		X	X	X	X

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. See above "Muscular Dystrophy Outcome Quality Measures" section for further details.

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.

The AAN worked with a medical librarian and did a supplementary web-based search to look for existing quality or performance measures for MD. Based upon the searches there are no existing quality measures for MDs. There is a strong need for valid and reliable quality of care measures for MD disorder management. Measures are also needed on the health plan level.

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

The AAN 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 MD 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/Retrospective 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

A denominator exclusion is a factor supported by the clinical evidence that removes a patient from inclusion in the measure population. For example, if the denominator indicates the measure is for all patients aged 0 to 18 years of age, a patient who is 19 years of age is excluded.

A denominator exception is a condition that should remove the patient, procedure or unit of measurement from the denominator only if the numerator criteria are not met. The AAN includes three possible types of exceptions for reasons why a patient should not be included in a measure denominator: medical, patient or system reasons.

- Medical exceptions may address:
  - Treatment, procedure, or measurement unit is not indicated (e.g., absence of organ/limb, already received/performed, etc.)
  -A contraindication (e.g., patient allergic history, potential adverse drug interaction, etc.)

- Patient exceptions may address:
  - Patient declinations
  - Cultural or religious beliefs

- System exception may address:
  - Resources limitations (e.g., particular vaccine was withdrawn from the market, transportation barriers, lack of appropriate specialty provider within a 500 mile radius, etc.)
  - Inability to pay for a test or intervention (i.e., payer-related limitations)

For each measure, there must be a clear rationale to permit an exception for a medical, patient, or system reason. The Work Group provided explicit exceptions when applicable for ease of use in eMeasure development.

Although this methodology does not require the external reporting of more detailed exception or exclusion data, the AAN requests that clinicians document the specific reasons for exception 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 clinician's exceptions or 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.

Testing and Implementation of the Measurement Set

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.

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

Numerator Statement	Patients prescribed appropriate DMD disease modifying pharmaceutical therapy*. *Current appropriate disease modifying pharmaceutical therapy for DMD: Corticosteroids
Denominator Statement	All patients diagnosed with Duchenne muscular dystrophy (DMD).
Denominator Exceptions	Exceptions: • Medication exception for not prescribing disease modifying pharmaceutical therapy (i.e., medical contraindication; patient already on corticosteroid; may not be medically appropriate depending upon functional capability, age, and existing risk factors) • Patient exception for not prescribing disease modifying pharmaceutical therapy (i.e., patient or family caregiver declines) • System exception for not prescribing disease modifying pharmaceutical therapy (i.e., 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 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 include 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 corticosteroids 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 and practice parameter guidelines, the panel strongly urges consideration of glucocorticosteroid therapy in all

patients who have DMD. (Formal Consensus Statement)[3] The goal of the use of glucocorticoids in the ambulatory child is the preservation of ambulation and the minimization of later respiratory, cardiac, and orthopedic complications, taking into account the well-described risks associated with chronic glucocorticoid administration. If such issues are pre-existing, the risk of side-effects might be increased. Particular care needs to be taken with such patients in deciding which glucocorticoid to choose, when to initiate treatment, and how best to monitor the child for any problems. A high index of suspicion for steroid-related side-effects needs to be maintained at all times. Prevention and management of side-effects needs to be proactive. Families should be provided with a steroid card or similar notification that the child is on steroids, listing emergency-care considerations in the setting of acute medical presentation, fracture, serious infection, need for surgery, or general anesthesia, to alert any medical professional with whom the child might come into contact.[3]

- Open discussion across the multidisciplinary team regarding the type and duration of specific interventions encourages transparency and shared decision-making. [4]
- Current recommendations indicate that the timing of initiation of glucocorticoid therapy must be individualized, considering the functional capabilities, age, and preexisting risk factors for adverse effects in each child. Initiation of glucocorticoids is not recommended for a child who is still gaining motor skills, which usually plateaus around the ages of 4-to-8 years. The child who takes longer to perform motor tasks in timed testing, loses a skill (such as climbing stairs), shows less endurance, or has more falls should be considered for starting glucocorticoid therapy.[5]
- Some variability about the dosing of glucocorticoids exists. The majority of available evidence indicates that glucocorticoids should be given in a single daily dose, commonly oral prednisone at a starting dose of 0.75 mg/kg/d. A recent study comparing daily prednisone (0.75mg/kg/d) versus high-dose (10 mg/kg/d) weekend prednisone demonstrated equal benefits and overall good tolerability of both dosing regimens. Another study compared daily doses of prednisone to alternate-day dosing; the results were that those boys assigned to the alternate-day therapy had significant loss of strength by 3 months compared to those on the daily dose regimen. Boys on the daily dose regimen did not lose strength for the duration of the study. Furthermore, no major differences in adverse effects were seen between the two groups.[10] If adverse effects such as weight gain require a decrease in dose, then a gradual tapering to dosages as low as 0.3 mg/kg/d may still be beneficial.[5]

[1]Mullender MG, Bom NA, De Kleuver M, et al. A Dutch Guideline for the Treatment of Scoliosis in Neuromuscular Disorders. Scoliosis 2008;3:14. Available at: http://www.scoliosisjournal.com/content/3/1/14 Accessed on February 15, 2014.
[2]Moxley RT, Ashwal S, Pandya S, et. Al. Practice Parameter: Corticosteroid Treatment of Duchenne Dystrophy: Report of the Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurology Society Neurology 2005 Jan 11;64(1):13-20. Reaffirmed February 2008.
[3]Bushby K, Finkel R, Birnkrant DJ, et al. Diagnosis and Management of Duchenne muscular dystrophy, part 1: diagnosis, and pharmacological and psychosocial management. Lancet Neurol 2010 Jan;9(1):77-93.
[4]Hull J, Aniapravan R, Chan E, et al. British Thoracic Society Guideline for Respiratory Management of Children With Neuromuscular Weakness. Thorax 2012;67: i1-i40
5. Gutierrez A, England JD. Administration of glucocorticoids in boys with Duchenne Muscular Dystrophy. Continuum 2013;19(6)1703-1708.

Rationale for the Measure

Gap in Care

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 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 DMD, 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 Muscular Dystrophy Association 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 orthopedic 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.

[1]Sejerson, T and Bushby K. Standards of care for Duchenne muscular dystrophy: brief TREAT-NMD recommendations. Adv Exp Med Biol 2009;652:13-21.
[2]Matthews DK, Adams KA, Miller LA. Use of corticosteroids in a population-based cohort of boys with Duchenne and Becker muscular dystrophy. J Child Neurol 2010; 25:1319 originally published online March 5, 2010.
[3]Griggs RC, Here BE, Reha A, et al. Corticosteroids in Duchenne muscular dystrophy: Major variations in practice. Muscle Nerve 2013; 48: 27–31.
[4]Bushby K, Finkel R, Birnkrant DJ, et al. Diagnosis and Management of Duchenne muscular dystrophy, part 1: diagnosis, and pharmacological and psychosocial management. Lancet Neurol 2010 Jan;9(1):77-93.
[5]Merlini L, Gennari L, Malaspina E, et al. Early corticosteroid treatment in 4 Duchenne muscular dystrophy patients: 14-year follow-up. Muscle and Nerve 2012; 45(6):796-802.
[6]Chamova T, Guergueltcheva V, Dimitrova T, et al. Corticosteroid treatment of patients with Duchenne muscular dystrophy-evaluation of their effect after the first year. Pediatriya 2012; 52(1):58-60.
[7]Moxley RT, Pandya S, Ciafaloni E, et al. Change in national history of Duchenne muscular dystrophy with long-term corticosteroid treatment: implications for management. J Child Neurol 2010; 25:1116. Originally published online June 25, 2010.

Measure Designation

Measure purpose	• Quality improvement • Accountability

Type of measure	• Process
Level of Measurement	• Individual practitioner
Care setting	• Outpatient visits • Nursing homes
Data source	• Electronic health record (EHR) data • Data registry

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 #2: MD Multidisciplinary Care Plan Developed or Updated
MUSCULAR DYSTROPHY

Measure Description

All patients diagnosed with a muscular dystrophy (MD) 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, dietician, 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
Denominator Statement	All patients diagnosed with a muscular dystrophy.
Denominator Exceptions	Exceptions: • Medical reason for not developing or updating a multidisciplinary care plan (i.e., plan was updated within 12 months of the date of the encounter) • Patient reason for not developing or updating a multidisciplinary care plan (i.e., patient or family caregiver declines) • System reason for not developing or reviewing a multidisciplinary care plan (i.e., 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 MD to neuromuscular centers to optimize the diagnostic evaluation and subsequent management. (Level B)[1,2] • L1. Clinicians should refer patients with MD 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 MD and other neuromuscular disorders in order to provide efficient and effective long-term care. (Level B)[1] • AA1. Clinicians caring for children with congenital muscular dystrophy (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 Duchenne muscular dystrophy (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)[3,4]

[1] Narayanaswami P, Weiss M, Selcen D, et al. Evidence-based Guideline: Diagnosis and Treatment of Limb-Girdle and Muscular Dystrophies. Report of the Guideline Development Subcommittee of the American Academy of Neurology and the PIRP of the American Association of Neuromuscular & Electrodiagnostic Medicine. UNDER DEVELOPMENT AND NOT YET APPROVED BY THE AAN BOARD OF DIRECTORS as of November 4, 2013.
[2] Kang PB, Morrison L, Iannaccone ST, et al. Evidence-based Guideline: Evaluation, Diagnosis and Management of Congenital Muscular Dystrophy. Report of the Guideline Development Subcommittee of the American Academy of Neurology and the PIRP of the American Association of Neuromuscular & Electrodiagnostic Medicine. UNDER DEVELOPMENT AND NOT YET APPROVED BY THE AAN BOARD OF DIRECTORS as of November 4, 2013.
[3] Bushby K, Finkel R, Birnkrant DJ, et al. Diagnosis and Management of Duchenne muscular dystrophy, part 1: diagnosis, and pharmacological and psychosocial management. Lancet Neurol 2010 Jan;9(1):77-93.
[4]Bushby K, Finkel R, Birnkrant DJ, et al. Diagnosis and Management of Duchene muscular dystrophy, part 2: implementation of multidisciplinary care. Lancet Neurol 2010; 9:177-89.

Rationale for the Measure

A systematic review of muscular dystrophies has highlighted the medical complexity of caring for patients with MD. 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.[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.[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, orthopedic management, psychosocial management, cardiac management, pulmonary management, GI/speech/swallowing/nutrition management, and corticosteroid management.[2]

One study pointed out disparities in receipt of healthcare and related services in adult men with Duchenne/Becker muscular dystrophy (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 MD 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 MD.[6]

[1] Narayanaswami P, Weiss M, Selcen D, et al. Evidence-based Guideline: Diagnosis and Treatment of Limb-Girdle and Muscular Dystrophies. Report of the Guideline Development Subcommittee of the American Academy of Neurology and the PIRP of the American Association of Neuromuscular & Electrodiagnostic Medicine. UNDER DEVELOPMENT AND NOT YET APPROVED BY THE AAN BOARD OF DIRECTORS as of November 4, 2013.
[2]Bushbya K, Bourkeb J, Bullock R, et al. The multidisciplinary management of Duchenne muscular dystrophy Current Paediatrics 2005; 15(4): 292–300.
[3]Bushby K, Finkel R, Birnkrant DJ, et al. Diagnosis and Management of Duchenne muscular dystrophy, part 1: diagnosis, and pharmacological and psychosocial management. Lancet Neurol 2010 Jan;9(1):77-93.
[4]Bushby K, Finkel R, Birnkrant DJ, et al. Diagnosis and Management of Duchene muscular dystrophy, part 2: implementation of multidisciplinary care. Lancet Neurol 2010; 9:177-89.
[5]Andrews JG, Davis MF, Meaney FJ. Correlates of care for young men with Duchenne and Becker muscular dystrophy. Muscle Nerve 2014 Jan;49(1):21-25. Available at: http://onlinelibrary.wiley.com/doi/10.1002/mus.23865/abstract Accessed November 5, 2013.
[6] Scully MA, et al. Can outcomes in Duchenne Muscular Dystrophy be improved by public reporting of data? Neurology 2013;80:583–589.

Measure Designation

Measure purpose	• Quality improvement • Accountability
Type of measure	• Process
Level of Measurement	• Individual practitioner
Care setting	• Inpatient Services • Outpatient visits • Nursing home
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.

Denominator (Eligible Population)

ICD-9 and ICD-10 Diagnosis Codes:

ICD-9 Code	ICD-10 Code
359 Muscular dystrophies and other myopathies	
359.0 Congenital hereditary muscular dystrophy	G71.2 Congenital myopathies
359.1 Hereditary progressive muscular dystrophy	G71.0 Muscular dystrophy
359.2 Myotonic disorders	
359.21 Myotonic muscular dystrophy	G71.11 Myotonic muscular dystrophy
359.22 Myotonia congenita	G71.12 Myotonia congenital
359.23 Myotonic chondrodystrophy	G71.13 Myotonic chondrodystrophy
359.8 Other myopathies	
359.89 Other myopathies	G72.89 Other specified myopathies
359.9 Myopathy, unspecified	G72.9 Myopathy, unspecified

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 #3: Evaluation of Pulmonary Status Ordered
MUSCULAR DYSTROPHY

Measure Description

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

Measure Components

Numerator Statement	Patients who had a pulmonary status evaluation* ordered. *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
Denominator Statement	All patients diagnosed with a muscular dystrophy.
Denominator Exceptions	Exceptions: • Medical exception for not ordering a pulmonary evaluation (i.e., patient cannot tolerate evaluation, MD phenotype does not warrant evaluation) • Patient exception for not ordering a pulmonary evaluation (i.e., patient or family caregiver declines an evaluation) • System exception for not ordering a pulmonary evaluation (i.e., clinic does not have the necessary equipment, patient cannot travel for testing, patient does not have insurance coverage)
Supporting Guideline & Other References	• Clinical assessment of respiratory health should be part of every medical consultation for children with neuromuscular weakness (NMW) and should be directed towards identifying progressive muscle weakness, ability to cope with respiratory infection, aspiration, progression of scoliosis and sleep-disordered breathing. [D][1] • Vital capacity should be measured in all patients with neuromuscular disease who are capable of performing spirometry as part of the respiratory assessment. [C][1] • C2. Clinicians should refer facioscapulohumeral muscular dystrophy (FSHD) patients with compromised pulmonary function studies (i.e., FVC < 60%) or with symptoms of excessive daytime somnolence or non-restorative sleep (e.g., frequent nocturnal arousals, morning headaches) for pulmonary or sleep medicine consultation for consideration of nocturnal sleep monitoring or nocturnal non-invasive ventilation to improve quality of life. (Level TBD)[2] • G3. Clinicians should refer MD patients with excessive daytime somnolence, non-restorative sleep (e.g., frequent nocturnal arousals, morning headaches, excessive daytime fatigue), or respiratory insufficiency based on PFTs for pulmonary or sleep medicine consultation for consideration of non-invasive ventilation to improve quality of life. (Level B)[2] • Obtain a detailed sleep history, evaluation of cough and airway protection, and serial measurements of FVC (sitting and supine) during routine office visits of patients with DM1 (Level A).[3] • C1. Clinicians should obtain baseline pulmonary function tests on all patients with FSHD. Patients with abnormal baseline pulmonary function tests or with any combination of severe proximal weakness, kyphoscoliosis, wheelchair dependence, or co-morbid conditions that may affect ventilation (e.g., COPD, cardiac disease) should be monitored with pulmonary function testing at every clinic visit.[2] • C3. All FSHD patients should have routine pulmonary function testing prior to all surgical procedures.[2]

- G1. In MD patients at time of diagnosis, or if they develop pulmonary symptoms later in their course, clinicians should order pulmonary function testing (spirometry along with maximal inspiratory/expiratory force in both the upright and if normal, supine positions) or refer for pulmonary evaluation to identify and treat respiratory insufficiency. (Level B)[4]
- G1a. In patients with a known high risk of respiratory failure (limb girdle muscular dystrophy (LGMD2I & MFM), clinicians should obtain periodic pulmonary function testing (spirometry along with maximal inspiratory/expiratory force in the upright position, and if normal, in the supine position) or evaluation by a pulmonologist to identify and treat respiratory insufficiency. (Level B)[4]
- G2. It is not obligatory to refer patients with LGMD2B and LGMD2L for pulmonary evaluation or pulmonary function testing unless symptomatic. (Level C)[4]
- D1b. Pulmonary function should be monitored in the awake and sleep states on a regular basis. (Level B)[4]
- Each child with confirmed Duchenne muscular dystrophy (DMD) should undergo an evaluation of respiratory status early (between ages 4 and 6), and tests of respiratory function should be performed at every clinic visit thereafter.[5]
- Care by a pulmonologist should be increased to every 3 to 6 months after the initiation of assisted ventilation or an airway clearance device.[5]
- Objective evaluation at each clinic visit should include: oxyhemoglobin saturation by pulse oximetry, spirometric measurements of FVC, FEV1, and maximal mid-expiratory flow rate, maximum inspiratory and expiratory pressures, and peak cough flow.[5]
- Awake carbon dioxide tension should be evaluated at least annually in conjunction with spirometry. Where available, capnography is ideal for this purpose. Arterial blood gas analysis is not necessary for routine follow-up of patients with DMD. If capnography is not available, then a venous or capillary blood sample should be obtained to assess for the presence of alveolar hypoventilation.[5]
- Additional measures of pulmonary function and gas exchange may be useful, including lung volumes, assisted cough peak flow, and maximum insufflation capacity.[5]
- Carefully evaluate patients for evidence of other respiratory disorders, such as obstructive sleep apnea, oropharyngeal aspiration, gastroesophageal reflux, and asthma.[5]
- All children with abnormal overnight oximetry should undergo more detailed sleep monitoring with at least oxycapnography. [1]
- When there is doubt about the cause of sleep disordered breathing, overnight polysomnography or sleep polysomnography should be performed. [1]
- Obtain a detailed sleep history, evaluation of cough and airway protection, and serial measurements of FVC (sitting and supine) during routine office visits of patients with DM1. (Level A)[3]
- Perform an overnight sleep study in patients with clinical complaints suggestive of sleep-related respiratory dysfunctions. (Level C)[3]
- Carefully evaluate patients for evidence of other respiratory disorders, such as obstructive sleep apnea, oropharyngeal aspiration, gastroesophageal reflux, and asthma.[5]
- Review sleep quality and symptoms of sleep-disordered breathing at every patient encounter.[6]
- In areas where full polysomnography is not readily available, overnight pulse oximetry with continuous CO2 monitoring provides useful information about nighttime gas exchange, although sleep-disordered breathing not associated with desaturation or CO2 retention will not be detected. A simple capillary blood gas

upon arousal in the morning can demonstrate CO2 retention, although not as sensitively as continuous capnography.[6]

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

[1]Hull J, Aniapravan R, Chan E, et al. British Thoracic Society Guideline for Respiratory Management of Children With Neuromuscular Weakness. Thorax 2012; 67: i1-i40.
[2] Tawil R, Kissel JT, Heatwole C, et al. Evidence-based Guideline: Evaluation, Diagnosis, and Management of Facioscapulohumeral Dystrophy. Report of the Guideline Development Subcommittee of the American Academy of Neurology and the PIRP of the American Association of Neuromuscular & Electrodiagnostic Medicine. UNDER DEVELOPMENT AND NOT YET APPROVED BY THE AAN BOARD OF DIRECTORS as of November 4, 2013.
[3] Ashizawa T, Moxley R, Day J, et al. Evidence-based Guideline: Evaluation and Management of Myotonic Dystrophy. Report of the Guideline Development Subcommittee of the American Academy of Neurology and the PIRP of the American Association of Neuromuscular & Electrodiagnostic Medicine. UNDER DEVELOPMENT AND NOT YET APPROVED BY THE AAN BOARD OF DIRECTORS as of November 4, 2013.
[4] Narayanaswami P, Weiss M, Selcen D, et al. Evidence-based Guideline: Diagnosis and Treatment of Limb-Girdle and Muscular Dystrophies. Report of the Guideline Development Subcommittee of the American Academy of Neurology and the PIRP of the American Association of Neuromuscular & Electrodiagnostic Medicine. UNDER DEVELOPMENT AND NOT YET APPROVED BY THE AAN BOARD OF DIRECTORS as of November 4, 2013.
[5]Wolfe L, N. Joyce N, C. McDonald C, et al. Management of Pulmonary Complications in Neuromuscular Disease. Phys Med Rehabil Clin N Am 2012; 23:829-853.
[6] 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. Reference is still up to date by Finder J. in 2009 in A 2009 Perspective on the 2004 American Thoracic Society Statement. Respiratory Care of the Patient with Duchenne Muscular Dystrophy. Pediatrics 2009;123:S239-S241.

Rationale for the Measure

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

Gap in care

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

Opportunity for Improvement

Patients with respiratory failure secondary to muscle weakness often have improved quality of life with noninvasive pulmonary ventilation.[3] Pulmonary function testing should therefore be done at regular intervals to identify the need for assistive respiratory equipment and initiate early noninvasive ventilation. Initiation of noninvasive ventilation can improve quality of life and prolong survival in patients with neuromuscular disease. Effective noninvasive strategies for management of hypoventilation, sleep-disordered breathing, and cough insufficiency are available for these patients.

	A respiratory action plan should be enacted with increasing disease severity. Therapeutic measures comprise airway clearance, respiratory muscle training, noninvasive nocturnal ventilation, daytime noninvasive ventilation, and continuous invasive ventilation. At the advanced stage of respiratory failure, attention should be paid to complications related to long-term mechanical ventilation, such as pneumothorax and tracheal hemorrhage. The American Thoracic Society (ATS) consensus statement on the respiratory care of patients with DMD has helped many patients receive improved care by offering clinicians guidance and helping medical directors of insurance companies make better decisions regarding use of technology to prevent morbidity and mortality.[3] However, there is considerable work remaining to aid patients with DMD or types of MD with pulmonary complications. [1]Kuru, S. Respiratory management in muscular dystrophies. Brain Nerve 2011;63(11):1229-36. [2]Katz SL, McKim D, Hoey L, et al. Respiratory management strategies for Duchenne muscular dystrophy: practice variation amongst Canadian sub-specialists. Pediatr Pulmonol 2013;48(1):59-66. [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. Reference is still up to date by Finder J. in 2009 in A 2009 Perspective on the 2004 American Thoracic Society Statement. Respiratory Care of the Patient with Duchenne Muscular Dystrophy. Pediatrics 2009;123:S239-S241.

Measure Designation

Measure purpose	• Quality improvement • Accountability
Type of measure	• Process
Level of Measurement	• Individual practitioner
Care setting	• Inpatient Services • 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.

Denominator (Eligible Population)

ICD-9 and ICD-10 Diagnosis Codes:

ICD-9 Code	ICD-10 Code
359 Muscular dystrophies and other myopathies	
359.0 Congenital hereditary muscular dystrophy	G71.2 Congenital myopathies
359.1 Hereditary progressive muscular dystrophy	G71.0 Muscular dystrophy

359.2 Myotonic disorders	
359.21 Myotonic muscular dystrophy	G71.11 Myotonic muscular dystrophy
359.22 Myotonia congenita	G71.12 Myotonia congenital
359.23 Myotonic chondrodystrophy	G71.13 Myotonic chondrodystrophy
359.8 Other myopathies	
359.89 Other myopathies	G72.89 Other specified myopathies
359.9 Myopathy, unspecified	G72.9 Myopathy, unspecified

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 #4: Evaluation of Cardiac Status Ordered
MUSCULAR DYSTROPHY

Measure Description

Patients diagnosed with a muscular dystrophy (MD) who had a cardiac status evaluation* ordered.

Measure Components

Numerator Statement	Patients who had a cardiac status evaluation ordered*. *Cardiac evaluation may include: referral for a consultation with a cardiologist, electrocardiograms, echocardiograms, and other rhythm monitoring such as Holter monitoring, cardiac imaging that are relevant to the patient's phenotype of MD.
Denominator Statement	All patients diagnosed with a muscular dystrophy.
Denominator Exceptions	Exceptions: • Medical exception for not ordering a cardiac evaluation (i.e., patient cannot tolerate the testing; MD phenotype is not associated with cardiac complications) • Patient exception for not ordering a cardiac evaluation (i.e., patient or family caregiver declines) • System reason for not ordering a cardiac evaluation (i.e., 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 facioscapulohumeral muscular dystrophy (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] • E1. Clinicians should refer newly diagnosed patients with limb girdle muscular dystrophy [LGMD]1A, LGMD1B, LGMD1D, LGMD1E, LGMD2C–K, LGMD2M–P, BMD, Emery-Dreifuss muscular dystrophy (EDMD), and MFM and MD patients without a specific genetic diagnosis for cardiology evaluation, including ECG and structural evaluation (echocardiography or cardiac MRI), even if they are asymptomatic from a cardiac standpoint, to guide appropriate management. (Level B)[3] • E1a. If ECG or structural cardiac evaluation (e.g., echocardiography) is abnormal, or if the patient has episodes of syncope, near-syncope, or palpitations, clinicians should order rhythm evaluation (e.g., Holter monitor or event monitor) to guide appropriate management. (Level B)[3] • E2. Clinicians should refer muscular dystrophy patients with palpitations or who are found to have symptomatic or asymptomatic tachycardia or arrhythmias for cardiology evaluation. (Level B)[3] • E3. Clinicians should refer MD patients with signs or symptoms of cardiac failure for cardiology evaluation (e.g., medical management, left ventricular assist device placement, or cardiac transplantation, as deemed necessary by the cardiologist) to prevent cardiac death. (Level B)[3]

- E4. It is not obligatory to refer or not to refer patients with LGMD2A, LGMD2B, and LGMD2L for cardiac evaluation, unless they develop overt cardiac signs or symptoms. (Level B) [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 Duchenne muscular dystrophy (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 Becker muscular dystrophy (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 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]

[1]Ashizawa T, Moxley R, Day J, et al. Evidence-based Guideline: Evaluation and Management of Myotonic Dystrophy. Report of the Guideline Development Subcommittee of the American Academy of Neurology and the PIRP of the American Association of Neuromuscular & Electrodiagnostic Medicine. UNDER DEVELOPMENT AND NOT YET APPROVED BY THE AAN BOARD OF DIRECTORS as of November 4, 2013.
[2]Tawil R, Kissel JT, Heatwole C, et al. Evidence-based Guideline: Evaluation, Diagnosis, and Management of Facioscapulohumeral Dystrophy. Report of the Guideline Development Subcommittee of the American Academy of Neurology and the PIRP of the American Association of Neuromuscular & Electrodiagnostic Medicine. UNDER DEVELOPMENT AND NOT YET APPROVED BY THE AAN BOARD OF DIRECTORS as of November 4, 2013.
[3] Narayanaswami P, Weiss M, Selcen D, et al. Evidence-based Guideline: Diagnosis and Treatment of Limb-Girdle and Muscular Dystrophies. Report of the Guideline Development Subcommittee of the American Academy of Neurology and the PIRP of the American Association of Neuromuscular & Electrodiagnostic Medicine. UNDER DEVELOPMENT AND NOT YET APPROVED BY THE AAN BOARD OF DIRECTORS as of November 4, 2013.
[4] Wolfe L, N. Joyce N, C. McDonald C, et al. Management of Pulmonary Complications in Neuromuscular Disease. Phys Med Rehabil Clin N Am 2012;23:829-853.
[5]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. Reference is still up to date by Finder J. in 2009 in A 2009 Perspective on the 2004 American Thoracic Society Statement. Respiratory Care of the Patient with Duchenne Muscular Dystrophy. Pediatrics 2009;123:S239-S241.
[6]American Academy of Pediatrics. Cardiovascular Health Supervision for Individuals Affected by Duchenne or Becker Muscular Dystrophy. Pediatrics 2005;116(6):1569-1573. A statement of reaffirmation for this policy was published on

Pediatrics 2008;123(2):1421.

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 DMD or BMD.[2] Such changes cause dilated cardiomyopathy in DMD, BMD and LGMD, cardiac arrhythmias in myotonic dystrophy, EDMD, LGMD and FSHD. Therefore, timely evaluation of cardiac status is important to prevent sudden 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.

[1]Narayanaswami P, Weiss M, Selcen D, et al. Evidence-based Guideline: Diagnosis and Treatment of Limb-Girdle and Muscular Dystrophies. Report of the Guideline Development Subcommittee of the American Academy of Neurology and the PIRP of the American Association of Neuromuscular & Electrodiagnostic Medicine. UNDER DEVELOPMENT AND NOT YET APPROVED BY THE AAN BOARD OF DIRECTORS as of November 4, 2013.
[2]American Academy of Pediatrics. Cardiovascular Health Supervision for Individuals Affected by Duchenne or Becker Muscular Dystrophy. Pediatrics 2005;116(6):1569-1573. A statement of reaffirmation for this policy was published on

	Pediatrics 2008;123(2):1421. [3] Bobo JK, Kenneson A, Kolor K, Brown MA. Adherence to American Academy of Pediatrics recommendations for cardiac care among female carriers of Duchenne and Becker muscular dystrophy. Pediatrics 2009;123(3): e471-e475. [4] Breton, R. and Mathieu, J. Usefulness of clinical and electrocardiographic data for predicting adverse cardiac events in patients with myotonic dystrophy. Can J Cardiol 2009;25(2):e23-e27.

Measure Designation

Measure purpose	• Quality improvement • Accountability
Type of measure	• Process
Level of Measurement	• Individual practitioner
Care setting	• Inpatient Services • 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.

Denominator (Eligible Population)

ICD-9 and ICD-10 Diagnosis Codes:

ICD-9 Code	ICD-10 Code
359 Muscular dystrophies and other myopathies	
359.0 Congenital hereditary muscular dystrophy	G71.2 Congenital myopathies
359.1 Hereditary progressive muscular dystrophy	G71.0 Muscular dystrophy
359.2 Myotonic disorders	
359.21 Myotonic muscular dystrophy	G71.11 Myotonic muscular dystrophy
359.22 Myotonia congenita	G71.12 Myotonia congenital
359.23 Myotonic chondrodystrophy	G71.13 Myotonic chondrodystrophy
359.8 Other myopathies	
359.89 Other myopathies	G72.89 Other specified myopathies
359.9 Myopathy, unspecified	G72.9 Myopathy, unspecified

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 #5: Scoliosis Evaluation Ordered
MUSCULAR DYSTROPHY

Measure Description

All visits for patients with a diagnosis of a muscular dystrophy (MD) 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.
Denominator Statement	All visits for patients with a diagnosis of a muscular dystrophy.
Denominator Exceptions	Exceptions: • Medical reason for not ordering a scoliosis evaluation (i.e., patient cannot tolerate evaluation, MD phenotype not associated with scoliosis) • Patient reason for not ordering a scoliosis evaluation (i.e., patient or family caregiver declines evaluation) • System reason for not ordering a scoliosis evaluation (i.e., patient has no insurance coverage for x-rays or referral for consultation evaluation)
Supporting Guideline & Other References	• I1. Clinicians should monitor MD patients for the development of spinal deformities to prevent resultant complications and preserve function. (Level B)[1] • I2. Clinicians should refer MD 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] • Wearing a rigid spinal bracing causes a reduction in both tidal ventilation and vital capacity in children with neuromusculardisease (evidence level 3). In boys with Duchenne muscular dystrophy (DMD), bracing may slow the progression ofscoliosis, but does not affect final scoliosis severity (evidence level 3).The effect of wearing a spinal brace on respiratory functionshould be assessed and weighed against the limited evidence of benefit in terms of affecting final scoliosis severity. [D] The primary consideration when planning surgery for childrenwith scoliosis associated with NMW should be comfort and quality of life.[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]

[1] Narayanaswami P, Weiss M, Selcen D, et al. Evidence-based Guideline: Diagnosis and Treatment of Limb-Girdle and Muscular Dystrophies. Report of the Guideline Development Subcommittee of the American Academy of Neurology and the PIRP of the American Association of Neuromuscular & Electrodiagnostic Medicine. UNDER DEVELOPMENT AND NOT YET APPROVED BY THE AAN BOARD OF DIRECTORS as of November 4, 2013.
[2]Hull J, Aniapravan R, Chan E, et al. British Thoracic Society Guideline for Respiratory Management of Children With Neuromuscular Weakness. Thorax 2012;67:i1-i40.
[3]Bushby K, Finkel R, Birnkrant DJ, et al. Diagnosis and Management of Duchene muscular dystrophy, part 2: implementation of multidisciplinary care. Lancet Neurol 2010;9:177-189.
[4] Mullender MG, Bom NA, De Kleuver M, et al. A Dutch Guideline for the Treatment of Scoliosis in Neuromuscular Disorders. Scoliosis 2008;3:14. Available at: http://www.scoliosisjournal.com/content/3/1/14 Accessed on February 15, 2014.

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.

[1] Narayanaswami P, Weiss M, Selcen D, et al. Evidence-based Guideline: Diagnosis and Treatment of Limb-Girdle and Muscular Dystrophies. Report of the Guideline Development Subcommittee of the American Academy of Neurology and the PIRP of the American Association of Neuromuscular & Electrodiagnostic Medicine. UNDER DEVELOPMENT AND NOT YET APPROVED BY THE AAN BOARD OF DIRECTORS as of November 4, 2013.
[2] Mullender MG, Bom NA, De Kleuver M, et al. A Dutch Guideline for the Treatment of Scoliosis in Neuromuscular Disorders. Scoliosis 2008;3:14. Available at: http://www.scoliosisjournal.com/content/3/1/14 Accessed on February 15, 2014.

Measure Designation	
Measure purpose	• Quality improvement • 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)

Denominator (Eligible Population)

ICD-9 and ICD-10 Diagnosis Codes:

ICD-9 Code	ICD-10 Code
359 Muscular dystrophies and other myopathies	
359.0 Congenital hereditary muscular dystrophy	G71.2 Congenital myopathies
359.1 Hereditary progressive muscular dystrophy	G71.0 Muscular dystrophy
359.2 Myotonic disorders	
359.21 Myotonic muscular dystrophy	G71.11 Myotonic muscular dystrophy
359.22 Myotonia congenital	G71.12 Myotonia congenital
359.23 Myotonic chondrodystrophy	G71.13 Myotonic chondrodystrophy
359.8 Other myopathies	
359.89 Other myopathies	G72.89 Other specified myopathies
359.9 Myopathy, unspecified	G72.9 Myopathy, unspecified

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)

MEASURE #6: Patient Referred for Physical, Occupational, or Speech/Swallowing Therapy
MUSCULAR DYSTROPHY

Measure Description

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

Measure Components

Numerator Statement	Patient visits where the patient was referred for physical, occupational, or speech/swallowing therapy.
Denominator Statement	All visits for patients diagnosed with a muscular dystrophy.
Denominator Exceptions	Exceptions: • Medical exception for not referring for physical, occupational, or speech/swallowing therapy. (i.e., patient does not need therapy based upon MD phenotype; patient already currently being seen by physical therapy (PT), occupational therapy (OT), or speech/swallowing specialist) • Patient exception for not referring for physical, occupational, or speech/swallowing therapy. (i.e., patient or family caregiver declines) • System exception for not referring for physical, occupational, or speech/swallowing therapy. (i.e., patient does not have insurance to cover therapy, patient cannot travel to the therapy facility)
Supporting Guideline & Other References	• L5. For patients with MD, 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, in order to preserve mobility and function and prevent contractures (Level B).[1] • L3. Clinicians should recommend that patients with MD have periodic assessments by a physical and occupational therapist for symptomatic and preventive screening (Level B)[1] • F1. Clinicians should refer MD 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)[1] • L1. Clinicians should refer patients with MD 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.] • H1. Physicians should consider referrals to allied health professionals, including physical, occupational, and speech therapists, seating and mobility specialists, rehabilitation specialists, and orthopedic surgeons to help maximize function and potentially slow the progression of musculoskeletal complications in children with congenial muscular dystrophy (CMD). (Level TBD)[2] • D3. Clinicians should encourage patients with facioscapulohumeral muscular dystrophy (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.[3]

- 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. [4]

[1] Narayanaswami P, Weiss M, Selcen D, et al. Evidence-based Guideline: Diagnosis and Treatment of Limb-Girdle and Muscular Dystrophies. Report of the Guideline Development Subcommittee of the American Academy of Neurology and the PIRP of the American Association of Neuromuscular & Electrodiagnostic Medicine. DRAFT. Not yet approved by the AAN Board of Directors. UNDER DEVELOPMENT AND NOT YET APPROVED BY THE AAN BOARD OF DIRECTORS as of November 4, 2013.
[2] Kang PB, Morrison L, Iannaccone ST, et al. Evidence-based Guideline: Evaluation, Diagnosis and Management of Congenital Muscular Dystrophy. Report of the Guideline Development Subcommittee of the American Academy of Neurology and the PIRP of the American Association of Neuromuscular & Electrodiagnostic Medicine. UNDER DEVELOPMENT AND NOT YET APPROVED BY THE AAN BOARD OF DIRECTORS as of November 4, 2013.
[3] Tawil R, Kissel JT, Heatwole C, et al. Evidence-based Guideline: Evaluation, Diagnosis, and Management of Facioscapulohumeral Dystrophy. Report of the Guideline Development Subcommittee of the American Academy of Neurology and the PIRP of the American Association of Neuromuscular & Electrodiagnostic Medicine. UNDER DEVELOPMENT AND NOT YET APPROVED BY THE AAN BOARD OF DIRECTORS as of November 4, 2013.
[4]Hull J, Aniapravan R, Chan E, et al. British Thoracic Society Guideline for Respiratory Management of Children With Neuromuscular Weakness. Thorax 2012;67:i1-i40.

Rationale for the Measure

Patients with MD 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 limb girdle muscular dystrophy (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 MD, 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 it 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
PT 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.

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.

[1]Narayanaswami P, Weiss M, Selcen D, et al. Evidence-based Guideline: Diagnosis and Treatment of Limb-Girdle and Muscular Dystrophies. Report of the Guideline Development Subcommittee of the American Academy of Neurology and the PIRP of the American Association of Neuromuscular & Electrodiagnostic Medicine. UNDER DEVELOPMENT AND NOT YET APPROVED BY THE AAN BOARD OF DIRECTORS as of November 4, 2013.
[3]Wang CH, Bonnemann CG, Rutkowski A, et al. Consensus Statement of Standards of Care for Congenital Muscular Dystrophies. J Child Neurol 2010;25:1559-1581. Originally published online November 15, 2010. Available at: http://jcn.sagepub.com/content/25/12/1559 Access on February 15, 2014.

Measure Designation

Measure purpose	• Quality improvement • Accountability
Type of measure	• Process

Level of Measurement	• Individual practitioner
Care setting	• Inpatient Consultations • Outpatient visits • Nursing Homes • Home Services • Rehabilitation Services

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.

Denominator (Eligible Population)

ICD-9 and ICD-10 Diagnosis Codes:

ICD-9 Code	ICD-10 Code
359 Muscular dystrophies and other myopathies	
359.0 Congenital hereditary muscular dystrophy	G71.2 Congenital myopathies
359.1 Hereditary progressive muscular dystrophy	G71.0 Muscular dystrophy
359.2 Myotonic disorders	
359.21 Myotonic muscular dystrophy	G71.11 Myotonic muscular dystrophy
359.22 Myotonia congenital	G71.12 Myotonia congenital
359.23 Myotonic chondrodystrophy	G71.13 Myotonic chondrodystrophy
359.8 Other myopathies	
359.89 Other myopathies	G72.89 Other specified myopathies
359.9 Myopathy, unspecified	G72.9 Myopathy, unspecified

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);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).
97001, 97002, 97003, 97004 (PT/OT evaluation)

MEASURE #7: Nutritional Status or Growth Trajectories Monitored
MUSCULAR DYSTROPHY

Measure Description

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

Measure Components

Numerator Statement	Patient visits where the patient's nutritional status or growth trajectories were monitored*. *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.
Denominator Statement	All visits for patients diagnosed with muscular dystrophy.
Denominator Exceptions	Exceptions: • Medical reason for not monitoring for nutrition or growth trajectory problems or referring for these purposes (i.e., patient is already being following by a nutritionist or other qualified specialist for these issues) • Patient reason for not monitoring for nutrition or growth trajectory problems or referring for these purposes (i.e., patient or family caregiver declines) • System reason for not monitoring for nutrition or growth trajectory problems or referring for these purposes (i.e., patient is unable to travel)
Supporting Guideline & Other References	• D2. The physician should refer the child with congenital muscular dystrophy (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 Duchenne muscular dystrophy (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. [O][3] • 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]

[1]Kang PB, Morrison L, Iannaccone ST, et al. Evidence-based Guideline: Evaluation, Diagnosis and Management of Congenital Muscular Dystrophy. Report of the Guideline Development Subcommittee of the American Academy of Neurology and the PIRP of the American Association of Neuromuscular & Electrodiagnostic Medicine. UNDER DEVELOPMENT AND NOT YET APPROVED BY THE AAN BOARD OF DIRECTORS as of November 4, 2013.
[2] Davidson Z, Truby H. A Review of Nutrition in Duchenne Muscular Dystrophy. J of Hum Nutri Diet 2009; 22(5):383-393.
[3]Hull J, Aniapravan R, Chan E, et al. British Thoracic Society Guideline for Respiratory Management of Children With Neuromuscular Weakness. Thorax 2012;67:i1-i40.
[4]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. Reference is still up to date by Finder J. in 2009 in A 2009 Perspective on the 2004 American Thoracic Society Statement. Respiratory Care of the Patient with Duchenne Muscular Dystrophy. Pediatrics 2009;123:S239-S241.

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 to commencing 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.[1]

Patients with MD 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.[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.[3,4] Overweight also has to be considered, particularly in the adult population because of the limited mobility of these patients.[3,4] Growth should be screened by regular weight measurements, completed by height or a surrogate height measurement (arm span or ulnar length).[3] Anticipator guidance and prevention of undernutrition/malnutrition and being overweight/obese should be goals from diagnosis throughout life.[4]

Opportunity for Improvement

Patients with CMD 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.[3]

[1] Davidson ZE, Truby H. A review of nutrition in Duchenne muscular dystrophy. J Hum Nutr Diet 2009;22(5):383-93.
[2] Narayanaswami P, Weiss M, Selcen D, et al. Evidence-based Guideline: Diagnosis and Treatment of Limb-Girdle and Muscular Dystrophies. Report of the Guideline Development Subcommittee of the American Academy of Neurology and the PIRP of the American Association of Neuromuscular & Electrodiagnostic Medicine. UNDER DEVELOPMENT AND NOT YET APPROVED BY THE AAN BOARD OF DIRECTORS as of November 4, 2013.

	[3]Wang CH, Bonnemann CG, Rutkowski A, et al. Consensus Statement of Standards of Care for Congenital Muscular Dystrophies. J Child Neurol 2010;25:1559-1581. Originally published online November 15, 2010. Available at: http://jcn.sagepub.com/content/25/12/1559 Access on February 15, 2014. [4]Bushby K, Finkel R, Birnkrant DJ, et al. Diagnosis and Management of Duchene muscular dystrophy, part 2: implementation of multidisciplinary care. Lancet Neurol 2010; 9:177-89.

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.

Denominator (Eligible Population)

ICD-9 and ICD-10 Diagnosis Codes:

ICD-9 Code	ICD-10 Code
359 Muscular dystrophies and other myopathies	
359.0 Congenital hereditary muscular dystrophy	G71.2 Congenital myopathies
359.1 Hereditary progressive muscular dystrophy	G71.0 Muscular dystrophy
359.2 Myotonic disorders	
359.21 Myotonic muscular dystrophy	G71.11 Myotonic muscular dystrophy
359.22 Myotonia congenital	G71.12 Myotonia congenital
359.23 Myotonic chondrodystrophy	G71.13 Myotonic chondrodystrophy
359.8 Other myopathies	
359.89 Other myopathies	G72.89 Other specified myopathies
359.9 Myopathy, unspecified	G72.9 Myopathy, unspecified

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).

MEASURE #8: Patient Queried about Pain and Pain Interference with Function
MUSCULAR DYSTROPHY

Measure Description

All visits for patients diagnosed with a muscular dystrophy (MD) 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 <u>not</u> limited to: • Numeric Rating Scale for Pain[1] • Faces Pain Scale[2] • Graded Chronic Pain Scale[3] • Visual Analogue Scale[4] • McGill Pain Questionnaire[5] • Short-Form McGill Pain Questionnaire[6] • Revised FLACC scale[7] • Individualized Numerical Rating Scale (INRS)[8] [1] Jensen MP. Pain assessment in clinical trials. In Wittink H, Carr D. eds., Pain Management: Evidence, outcomes, and quality of life in pain treatment. 1st ed. Amsterdam: Elsevier. 2008:57-88. [2]Bieri D, Reeve RA, Champion GD, et al. The Faces Pain Scale for the self-assessment of the severity of pain experienced by children: development, initial validation, and preliminary investigation for ratio scale properties. Pain. 1990;41:139–150. [3]Von Korff M, Ormel J, Keefe FJ, Dworkin SF. Grading the severity of chronic pain. Pain 1992;50:133-149. [4]Carlsson AM. Assessment of chronic pain. Aspects of the reliability and validity of the visual analogue scale. Pain 1983;16(1):87-101. [5] Melzack R. The McGill Pain Questionnaire: Major properties and scoring methods. Pain.1975;1(3):227-299. [6] Melzack R. The short-form McGill pain questionnaire. Pain 1987;30(2):191-197. [7] Malviya S, Voepel-Lewis T, Burke C, et al. The revised FLACC observational pain tool: improved reliability and validity for pain assessment in children with cognitive impairment. Paediatr Anaesth 2006;16(3):258-65. [8] Solodiuk JC, Scott-Sutherland J, Meyers M, et al. Validation of the Individualized Numeric Rating Scale (INRS): a pain assessment tool for nonverbal children with intellectual disability. Pain 2010;150(2):231- 36.
Denominator Statement	All visits for patients diagnosed with a muscular dystrophy.
Denominator Exceptions	Exceptions: • Patient reason for not querying about pain and pain interference with function (i.e., 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 facioscapulohumeral muscular dystrophy (FSHD).[2] [1]Hull J, Aniapravan R, Chan E, et al. British Thoracic Society Guideline for Respiratory Management of Children With Neuromuscular Weakness. Thorax 2012;67:i1-i40. [2] Tawil R, Kissel JT, Heatwole C, et al. Evidence-based Guideline: Evaluation, Diagnosis, and Management of Facioscapulohumeral Dystrophy. Report of the Guideline Development Subcommittee of the American Academy of Neurology and the PIRP of the American Association of Neuromuscular & Electrodiagnostic Medicine. UNDER DEVELOPMENT AND NOT YET APPROVED BY THE AAN BOARD OF DIRECTORS as of November 4, 2013.

	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 limb girdle muscular dystrophies (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. [1]Jensen MP, Hoffman AJ, Stoelb BL, et al. Chronic pain in persons with myotonic dystrophy and facioscapulohumeral dystrophy. Arch Phys Med Rehabil. 2008;89(2):320-8. [2] Miro J, Gertz KJ, Carter GT, Jensen MP. Chronic pain in neuromuscular disease. Pain site and intensity differentially impacts function. Phys Med Rehabil Clin N Am 2012;23:895-902. [3]Narayanaswami P, Weiss M, Selcen D, et al. Evidence-based Guideline: Diagnosis and Treatment of Limb-Girdle and Muscular Dystrophies. Report of the Guideline Development Subcommittee of the American Academy of Neurology and the PIRP of the American Association of Neuromuscular & Electrodiagnostic Medicine. UNDER DEVELOPMENT AND NOT YET APPROVED BY THE AAN BOARD OF DIRECTORS as of November 4, 2013. [4]Alschuler KN, Jensen MP, Goetz MC, et al. Effects of pain and fatigue on physical functioning and depression in persons with muscular dystrophy. Disability and Health J 2012;5(4):277–283. [5]Wang CH, Bonnemann CG, Rutkowski A, et al. Consensus Statement of Standards of Care for Congenital Muscular Dystrophies. J Child Neurol 2010;25:1559-1581. Originally published online November 15, 2010. Available at: http://jcn.sagepub.com/content/25/12/1559 Access on February 15, 2014. [6]Engel JM, Kartin D, Carter GT, et al. Pain in youths with neuromuscular disease. Am J Hosp Palliat Care 2009;26:405-412. [7]Tiffreau V, Viet G, Thevenon A. Pain and neuromuscular disease: the results of a survey. Am J. Phys Med Rehabil. 2006;85:756-766.

Measure Designation	
Measure purpose	• Quality improvement • Accountability
Type of measure	• Process
Level of Measurement	• Individual practitioner
Care setting	• Outpatient visits • Nursing homes • Home services • Rehabilitation
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.

Denominator (Eligible Population)

ICD-9 and ICD-10 Diagnosis Codes:

ICD-9 Code	ICD-10 Code
359 Muscular dystrophies and other myopathies	
359.0 Congenital hereditary muscular dystrophy	G71.2 Congenital myopathies
359.1 Hereditary progressive muscular dystrophy	G71.0 Muscular dystrophy
359.2 Myotonic disorders	
359.21 Myotonic muscular dystrophy	G71.11 Myotonic muscular dystrophy
359.22 Myotonia congenital	G71.12 Myotonia congenital
359.23 Myotonic chondrodystrophy	G71.13 Myotonic chondrodystrophy
359.8 Other myopathies	
359.89 Other myopathies	G72.89 Other specified myopathies
359.9 Myopathy, unspecified	G72.9 Myopathy, unspecified

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);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).
97001, 97002, 97003, 97004 (PT/OT evaluation)

MEASURE #9: Patient Counseled About Advanced Health Care Decision-Making, Palliative Care, or End-Of-Life Issues

MUSCULAR DYSTROPHY

Measure Description

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

Measure Components

Numerator Statement	Patients or caregivers who were counseled about advanced health care decision-making, palliative care, or end-of-life issues* at least once annually. *Advanced health care decision making, palliative care and end-of-life issues may include: emotional, spiritual, developmental, or physical dimensions.
Denominator Statement	All patients with a diagnosis of a muscular dystrophy.
Denominator Exceptions	Exceptions: • Medical exception for not counseling about advanced health care decision making, palliative care or end-of-life issues (i.e., patient is unable to communicate and caregiver is not available; not indicated because of early stage of disease without any comorbid complications)
Supporting Guideline & Other References	• L4. While respecting and protecting patient autonomy, clinicians should proactively anticipate and facilitate patient and family decision making as the disease progresses, including decisions regarding 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. [2] • Pediatric palliative care principles are directly applicable to the context of pediatric neuromuscular disease because the focus is on 'survivorship' and long-term multidimensional support aimed at maintaining quality of life. In neuromuscular disease management, a framework has developed that integrates the multidimensional approach of palliative care with curative interventions; focus is on what can be offered, rather than what cannot be done. Active palliative care in this context anticipates crucial milestones, which may precipitate vulnerabilities and offers flexible re-evaluation of goals of care in line with prognosis.Children's hospices in the UK have developed an important role with regard to children and young people with neuromuscular disease offering a wide range of supportive services unavailable elsewhere. Many expert review papers state the importance of integrating palliative care services into the mainstream care of children and young people with neuromuscular disease; more objective studies are needed to endorse the benefits of this approach. Evidence: Older studies suggest that the palliative care model has much to offer individuals with progressive neuromuscular conditions and their families. Children and young people with neuromuscular disease, especially those with Duchenne muscular dystrophy (DMD), form a large proportion of the cases cared for by the children's hospices in the UK. Most support is multidimensional. Planned stays allow access to peer support and social activities that are often curtailed or restricted in the wider community as a consequence of disability.

Referrals peak for DMD in adolescence at a time when those with DMD are losing ambulation and when patients often have greater physical and emotional needs. Recommendation: Families need access to skilled experts for multidimensional coordinated palliative care support, providing regular review of their needs at various stages in their condition. Good practice point: Generic palliative care skills should be cascaded to otherprofessionals providing neuromuscular services.[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. [2]
- Assisting patients, parents and caregivers to make informed choices that are consistent with their own values and preferences requires physicians to engage patients and their parents and caregivers in a process of mutual participation in decision-making with full disclosure of all information in a sensitive and timely fashion [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 [2]
- Patients and families need to have ownership of the advance care plan and be educated as to its uses. [2]
- Advance care plans should be reviewed by the multidisciplinary team on a regular basis. [2]
- Families need access to skilled experts for multidimensional coordinated palliative care support, providing regular review of their needs at various stages in their condition.[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]

[1] Narayanaswami P, Weiss M, Selcen D, et al. Evidence-based Guideline: Diagnosis and Treatment of Limb-Girdle and Muscular Dystrophies. Report of the Guideline Development Subcommittee of the American Academy of Neurology and the PIRP of the American Association of Neuromuscular & Electrodiagnostic Medicine. UNDER DEVELOPMENT AND NOT YET APPROVED BY THE AAN BOARD OF DIRECTORS as of November 4, 2013.

[2]Hull J, Aniapravan R, Chan E, et al. British Thoracic Society Guideline for Respiratory Management of Children With Neuromuscular Weakness. Thorax 2012;67:i1-i40.

[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. Reference is still up to date by Finder J. in 2009 in A 2009 Perspective on the 2004 American Thoracic Society Statement. Respiratory Care of the Patient with Duchenne Muscular Dystrophy. Pediatrics 2009;123:S239-S241.
[4]McKim D, Road J, Avendano M, et al. Home Mechanical Ventilation: A Canadian Thoracic Society Clinical Practice Guideline. Can Resp J 2011;18(4):197-215.

Rationale for the Measure
An important aspect of ongoing management includes proactively preparing patients with MD 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 MD 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 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 advance care planning.[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 advanced care planning.

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. [4]

Opportunity for Improvement
Health care providers should educate patients and families that palliative care is complementary to care with curative intent and that incorporating 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 a "life-limiting" diagnosis and a "life-threatening" episode, considering that 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.[5]

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.[5]

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.[5] [1] Narayanaswami P, Weiss M, Selcen D, et al. Evidence-based Guideline: Diagnosis and Treatment of Limb-Girdle and Muscular Dystrophies. Report of the Guideline Development Subcommittee of the American Academy of Neurology and the PIRP of the American Association of Neuromuscular & Electrodiagnostic Medicine. UNDER DEVELOPMENT AND NOT YET APPROVED BY THE AAN BOARD OF DIRECTORS as of November 4, 2013. [2]Carter GT, Joyce NC, Abresch AL, et al. Using palliative care in progressive neuromuscular disease to maximize quality of life. Phys Med Rehabil Clin N Am 2012;23:903-909. [3]Edwards JD, Kun SS, Grahan RJ, Keens TG. End-of-Life Discussions and Advance Care Planning for Children on Long-Term Assisted Ventilation with Life-Limiting Conditions. J Palliat Care. 2012;28(1):21–27. [4]Arias R, Andrews J, Pandya S, et al. Palliative care services in families of males with Duchenne muscular dystrophy Muscle & Nerve 2011;44:93–101. [5]Wang CH, Bonnemann CG, Rutkowski A, et al. Consensus Statement of Standards of Care for Congenital Muscular Dystrophies. J Child Neurol 2010;25:1559-1581. Originally published online November 15, 2010. Available at: http://jcn.sagepub.com/content/25/12/1559 Access on February 15, 2014.

Measure Designation

Measure purpose	• Quality improvement • Accountability
Type of measure	• Process
Level of Measurement	• Individual practitioner
Care setting	• Inpatient consultation • 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.

Denominator (Eligible Population)

ICD-9 and ICD-10 Diagnosis Codes:

ICD-9 Code	ICD-10 Code
359 Muscular dystrophies and other myopathies	
359.0 Congenital hereditary muscular dystrophy	G71.2 Congenital myopathies
359.1 Hereditary progressive muscular dystrophy	G71.0 Muscular dystrophy
359.2 Myotonic disorders	

359.21 Myotonic muscular dystrophy	G71.11 Myotonic muscular dystrophy
359.22 Myotonia congenita	G71.12 Myotonia congenital
359.23 Myotonic chondrodystrophy	G71.13 Myotonic chondrodystrophy
359.8 Other myopathies	
359.89 Other myopathies	G72.89 Other specified myopathies
359.9 Myopathy, unspecified	G72.9 Myopathy, unspecified

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);
99307, 99308, 99309, 99310 (Subsequent Nursing Facility Care);
99319 (Other Nursing Facility Services);
99341, 99342, 99343, 99344, 99345 (Home Services-New Patient);
99347, 99348, 99349, 99350 (Home Services-Established Patient).
97001, 97002, 97003, 97004 (PT/OT evaluation)

Contact Information

American Academy of Neurology
201 Chicago Avenue
Minneapolis, MN 55415
(612) 928-6100
quality@aan.com