**Measure Description**

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

**Measure Components**

<table>
<thead>
<tr>
<th>Numerator Statement</th>
<th>Patients who had a cardiac status evaluation ordered*.</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>*Cardiac evaluation may include: referral for a consultation with a cardiologist, electrocardiograms, echocardiograms, and other rhythm monitoring such as Holter monitoring, cardiac imaging that are relevant to the patient’s phenotype of MD.</td>
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</tbody>
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<table>
<thead>
<tr>
<th>Denominator Statement</th>
<th>All patients diagnosed with a muscular dystrophy.</th>
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</table>

**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)

- 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)  

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)  

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.  

All individuals with Duchenne muscular dystrophy (DMD) require regular cardiac evaluation with annual electrocardiograms and echocardiograms, starting at least by school age.  

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.)  

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.)  

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.)  

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.)  

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

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.)  

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.)

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 death due to arrhythmias, morbidity due to cardiomyopathy and resultant congestive heart failure and to improve outcome.

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

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

<table>
<thead>
<tr>
<th>Measure Designation</th>
<th>ICD-9 and ICD-10 Diagnosis Codes:</th>
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<tr>
<td>Measure purpose</td>
<td>ICD-9 Code</td>
</tr>
<tr>
<td>Quality improvement</td>
<td>359 Muscular dystrophies and other myopathies</td>
</tr>
<tr>
<td>Accountability</td>
<td>359.0 Congenital hereditary muscular dystrophy</td>
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<tr>
<td></td>
<td>359.1 Hereditary progressive muscular dystrophy</td>
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<td></td>
<td>359.2 Myotonic disorders</td>
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<td></td>
<td>359.21 Myotonic muscular dystrophy</td>
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<tr>
<td></td>
<td>359.22 Myotonia congenita</td>
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<tr>
<td></td>
<td>359.8 Other myopathies</td>
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<tr>
<td></td>
<td>359.89 Other myopathies</td>
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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:

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<thead>
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<tbody>
<tr>
<td>359 Muscular dystrophies and other myopathies</td>
<td>G71.2 Congenital myopathies</td>
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<tr>
<td>359.0 Congenital hereditary muscular dystrophy</td>
<td>G71.0 Muscular dystrophy</td>
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<td>359.1 Hereditary progressive muscular dystrophy</td>
<td>G71.11 Myotonic muscular dystrophy</td>
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<td>359.2 Myotonic disorders</td>
<td>G71.12 Myotonia congenita</td>
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<td>359.21 Myotonic muscular dystrophy</td>
<td>G71.13 Myotonic chondrodystrophy</td>
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<tr>
<td>359.22 Myotonia congenita</td>
<td>G71.89 Other specified myopathies</td>
</tr>
<tr>
<td>359.8 Other myopathies</td>
<td>G72.9 Myopathy, unspecified</td>
</tr>
<tr>
<td>359.89 Other myopathies</td>
<td>G72.9 Myopathy, unspecified</td>
</tr>
</tbody>
</table>

**AND**

CPT E/M Service Code:
- 99221, 99222, 99223 (Initial hospital care)
- 99231, 99232, 99233 (Subsequent hospital care)
- 99201, 99202, 99203, 99204, 99205 (Office or other outpatient visit-New Patient)

99211, 99212, 99213, 99214, 99215 (Office or other outpatient visit-Established Patient);
99241, 99242, 99243, 99244, 99245 (Office or Other Outpatient Consultation-New or Established Patient);
99304, 99305, 99306 (Initial nursing facility care, per day)
99307, 99308, 99309, 99310 (Subsequent nursing facility care, per day)
97001, 97002, 97003, 97004 (PT/OT evaluation)
99324, 99325, 99326, 99327, 99328 (Domiciliary visit, new patient)
99334, 99335, 99336, 99337 (Domiciliary visit, established patient)
99341, 99342, 99343, 99344, 99345 (Home visit, new patient)
99347, 99348, 99349, 99350 (Home visit, established patient)