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**

<table>
<thead>
<tr>
<th>Numerator Statement</th>
<th>Patients or caregivers who were counseled about advanced health care decision-making, palliative care, or end-of-life issues* at least once annually.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Denominator Statement</td>
<td>All patients with a diagnosis of a muscular dystrophy.</td>
</tr>
</tbody>
</table>

**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)
- 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.

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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 other professionals 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


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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. Palliative care is useful to alleviate the suffering of these patients.

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. 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. 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; pastoral care (27%), respite care (18%), pain management (12%), and hospice care (6%). Only 8 respondents (25%) reported having any type of directive document in place.

Opportunity for Improvement

Health care providers should educate patients and families that palliative care is complementary to care with curative intent and that 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 the trajectory of life toward death will be highly variable and certainly individual. Incorporating palliative care from diagnosis can benefit the patient, family, and medical team as they anticipate and make decisions regarding interventions that affect both the duration and quality of these individuals' lives.

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

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


**Measure Designation**

| Measure purpose   | Quality improvement  
<table>
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<tbody>
<tr>
<td></td>
<td>Accountability</td>
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</table>

**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.

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

**AND**

CPT E/M Service Code:
- 99221, 99222, 99223 (Initial hospital care)
- 99231, 99232, 99233 (Subsequent hospital care)
- 99201, 99202, 99203, 99204, 99205 (Office or other outpatient visit-New Patient)
- 99211, 99212, 99213, 99214, 99215 (Office or other outpatient visit-Established Patient)
- 99241, 99242, 99243, 99244, 99245 (Office or Other Outpatient Consultation-New or Established Patient)
- 99304, 99305, 99306 (Initial Nursing Facility Care)
- 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)