American Academy of Neurology

Amyotrophic Lateral Sclerosis

Performance Measurement Set

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

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

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Executive Summary: Towards Improving Outcomes for Patients with ALS

The American Academy of Neurology (AAN) formed an Amyotrophic Lateral Sclerosis Work Group to identify and define quality measures towards improving outcomes for patients with Amyotrophic Lateral Sclerosis (ALS).

The work group focused on quality measures that would be applicable to patients with an established diagnosis of ALS. The measures are focused mostly on the underutilization of aspects of evaluation and management of ALS.

Reasons for Prioritizing Improvement in Amyotrophic Lateral Sclerosis

High Impact Topic Area

- ALS, also known as Lou Gehrig's disease is a type of motor neuron disease that is a rapidly progressive and fatal neurological disease.[1] 20-30,000 people in the US have ALS.[1] 5,000 people are diagnosed with ALS in the US annually.[1] No cure exists for ALS. Newer pharmacotherapy agents have been found to reduce the progression, but not halt the disease development.[1]
- The prevalence of ALS is said to be between six and eight cases per 100,000 in the population. Using the higher prevalence estimate and data from the 2000 U.S. census, nearly 22,600 Americans are living with ALS at any one time. Since ALS is a disease of aging, as the U.S. population increases and ages, an increase in the prevalence of ALS can be anticipated.[2]
- Cognitive dysfunction is seen in 20–50%, and 3–5% develop dementia that is usually of frontotemporal type.[3]
- The lifetime risk for developing ALS for individuals aged 18 years of age and older has been estimated to be 1 in 350 for men and 1 in 420 for women[5b] ,with male sex, increasing age and hereditary disposition being the main risk factors.[6]
- Most patients with ALS die within 2 to 5 years of onset.[7] Only 10% of ALS patients survive for 10 years or more.[8]
- Pseudobulbar affect (PBA), excessive laughing or crying, or involuntary emotional expression disorder affects 20%–50% of patients with ALS, especially in pseudobulbar palsy.[9] Patients are embarrassed and isolated by these symptoms, which in turn greatly diminishes the patients' quality of life.
- Sialorrhea, or drooling, is embarrassing, socially isolating, and is associated with aspiration pneumonia. The prevalence is estimated at 50%, and 70% of patients receiving oral medications for treatment reported benefit (Class III).[10,20]
- Fatigue may be a symptom of depression, poor sleep, abnormal muscle activation, immobility, or respiratory dysfunction. Fatigue diminishes quality of life for patients with ALS. Fatigue was a side effect of therapy in 26% of patients taking riluzole vs. 13% taking placebo.[21] Asthenia occurred in 18% of patients taking riluzole vs. 12% of patients taking placebo in a larger study.[22]
- ALS patients have dysarthria in nearly all patients with bulbar onset and nearly 40% of ALS patients with spinal onset. More than 95% of ALS patients cannot speak before death and patients who accept gastrostomy tube, non-invasive ventilation or tracheostomy-ventilation have a greater need for augmentative alternative communication as the disease progresses.[32-35]

Demonstrated Opportunity for Improvement

- Treatments for ALS are underutilized even in specialized clinics.[56,57]
- Riluzole is currently the only available disease modifying pharmacotherapy available to slow down progression of ALS. Only 60% of patients are taking the riluzole in the United States, compared to nearly 100% in European countries (France, Italy, Germany).[57]
- There is now considerable evidence for cognitive and behavioral manifestations in ALS. The domain of cognitive and behavioral impairment in ALS is a rapidly evolving field and there is little consensus regarding diagnostic criteria and assessment methods.[59]
- Treatment for both refractory sialorrhea and pseudobulbar affect are underutilized in ALS.[56,57] Many practitioners are unaware of the condition and also of the recently approved treatment for pseudobulbar

affect.[67] Most other symptoms of ALS are treatable, albeit with less evidence base. Still, most symptoms in ALS can be treated and studies suggest that these treatments are underutilized.[56,57]

- Survival is improved with use of noninvasive ventilation. Utilization rates increased from 16% of ALS patients with respiratory insufficiency to 51%[71,72]. Over 65% of ALS patients have said that they would want respiratory support indicating that noninvasive ventilation is still underutilized in ALS patients.[56,57, 72] Even though published guidelines to date have made a positive impact by increasing utilization rates, underutilization of this important treatment is still evident.[56,57]
- Not all patients who should have enteral feeding receive this treatment with only 43% of ALS patients who met guideline indications for gastrostomy tube placement undergoing the procedure. [57] Increasing proportions of patients who should receive this treatment are now receiving treatment but not all. [56, 72,76,77]
- Speech assessment in ALS patients identifies dysarthria, independent of dysphagia that limits communication and maintenance of their active communicator role. Speech correction should focus on the maintenance of functional communication. Assessment should involve measurement of speech rate (words per minute). Augmentative communication is recommended when speech rate is less than 125 words per minute. Nearly 88% of ALS patients are evaluated by this criterion, but fewer than half implement appropriate interventions. [78-82]
- Patients with neurologic or general conditions associated with an increased risk of falling should be asked about recent falls and further examined for the presence of specific neurologic deficits that predict falls. If substantial risks of falls are identified, appropriate interventions that are described in other evidence-based guidelines may be considered.[86] There is an increased prevalence of falls in ALS patients of 0.7 falls per patient per 6 months.[88]

Disparities

- All races and ethnic backgrounds are affected by ALS.[1]
- ALS most common in individuals 40-60 years old, but younger and older people can develop the disease.[1]
- Men are more likely to develop ALS than women. Studies suggest an overall ratio of about 1.5 men to every woman who develops ALS in Western countries.[2]

Rigorous Clinical Evidence Base

Evidence-based clinical practice guidelines and consensus papers are available for the management of Amyotrophic Lateral Sclerosis. This measurement set is based upon guidelines or consensus papers from:

- American Academy of Neurology
- European Federation of Neurological Sciences
- British Thoracic Society/Association of Chartered Physiotherapists in Respiratory Care (Bott, J, Blumenthal S, Buxton M, et al. Guidelines for the physiotherapy management of the adult, medical, spontaneously breathing patient. *Thorax* 2009; 64:1-51.)
- Systematic Reviews: Heffernan C, Jenkinson C, Holmes T, et al. C. Management of respiration in MND/ALS patients: An evidence based review. *Amyotrophic Lateral Sclerosis* 2006; 7(1):5-15. Tripodoro VA, De Vito EL. Management of dyspnea in advanced motor neuron diseases. *Curr Opin Support Palliat Care* 2008;2(3):173-9.
- Cochrane Review: Miller RG, Mitchell JD, Lyon M, More DH. Riluzole for amyotrophic lateral sclerosis (ALS)/motor neuron disease (MND) (Review). 2009 The Cochrane Library. 2:1-27.

Amyotrophic Lateral Sclerosis Outcomes

The work group attempted to develop measures of outcomes along with measures of processes that may improve patient outcomes for ALS patients. The Work Group decided to focus on performance measures based upon processes that may achieve desired outcomes and reflect high quality care.

Desired outcomes for ALS include:

1. Care coordination between multiple providers
2. Enhance patient quality of life

3. Slowing ALS disease progression
4. Minimize ALS symptoms
5. Educate patients and promote patient-centered decision making
6. Avoid emergent decision making/promote advanced planning
7. Reduce hospitalizations

Amyotrophic Lateral Sclerosis Work Group Recommendations:

Process measures: Several processes of care demonstrated to improvement outcomes for patient with Amyotrophic Lateral Sclerosis are recommended:

Amyotrophic Lateral Sclerosis Measures
Measures addressing accurate and appropriate evaluation/monitoring of disease status and associated symptoms to guide treatment options
Measure #1: ALS Multidisciplinary Care Plan Developed or Updated
Measure #3: ALS Cognitive and Behavioral Impairment Screening
Measure #4: ALS Symptomatic Therapy Treatment Offered
Measure #5: ALS Respiratory Insufficiency Querying and Referral for Pulmonary Function Testing
Measure #7: ALS Screening for Dysphagia, Weight Loss and Impaired Nutrition
Measure #8: ALS Nutritional Support Offered
Measure #9: ALS Communication Support Referral
Measures addressing effective therapeutic options in eligible patients
Measure #2: Disease Modifying Pharmacotherapy for ALS Discussed
Measure #6: ALS Noninvasive Ventilation Treatment for Respiratory Insufficiency Discussed
Measures addressing increasing patient awareness of advanced planning
Measure #10: ALS End of Life Planning Assistance
Measures addressing patient safety
Measure #11: ALS Falls Querying*
Amyotrophic Lateral Sclerosis Measures

These measures are designed for individual practitioner level quality improvement. Unless otherwise indicated the measures are appropriate for accountability if the appropriate methodological, statistical, and implementation rules are followed. *This measure is for quality initiatives only.

Other Potential Measures

The Work Group considered several other potential measures, though ultimately decided they were not appropriate for inclusion in the measurement set.

Measure Harmonization

When existing measures are available for the same measurement topic, the AAN attempts to harmonize the measures to the extent it is feasible. The AAN works to ensure there is no duplication of existing measures. However, the one palliative care measure from the American Geriatrics Society is limited to those over the age of 65. The work group felt it was important to not exclude patients under 65 years old who have ALS from an end of life planning measure. In addition, the Institute for Clinical Systems Improvement (ICSI) that refers to a palliative care plan for those with debilitating disease. However, this measure does not reference specific end of life needs

that are relevant for patients with ALS. This measure was also not developed by a medical specialty society and the methods used to develop the measure are unclear.
For the 2012, Physician Quality Reporting System (PQRS) there are three measures that relate to falls. PQRS #154 is a Falls Risk Assessment, #155 is about a plan of care, and #318 is screening for future fall risk. However, all three refer to only patients 65 years and older, and many ALS patients will not fit the denominator, so therefore, a measure specific to ALS would not be overlapping.

Existing Quality Improvement (QI) Initiative or Collaborative for Measure Implementation
The American Academy of Neurology has developed a performance in practice program for maintenance of certification (MOC), NeuroPI[18], which meets the American Board of Psychiatry and Neurology (ABPN) requirements for MOC Performance in Practice requirements. The NeuroPI will contain a new module for Amyotrophic Lateral Sclerosis based upon the measures developed in this measurement set. The measures will be used as the basis of the module content. A separate formal testing effort for reliability and validity is also planned for this measurement set.

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

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

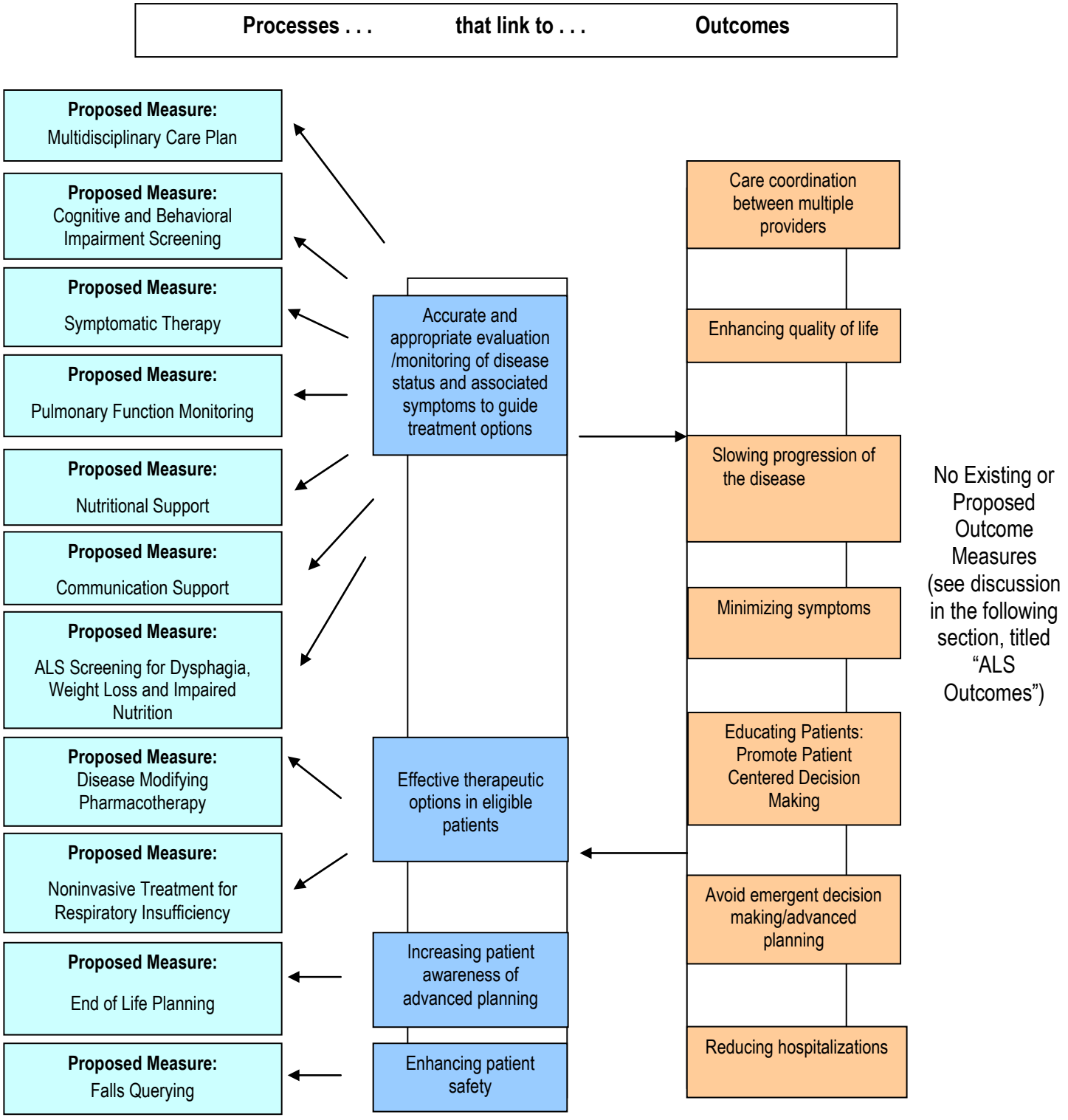
Because administrative claims are currently the only available sources of data, specifications to collect and report on the Amyotrophic Lateral Sclerosis measures for administrative claims are included in this document.

The AAN is in the process of creating data elements required for electronic capture with Electronic Health Records (EHRs). A listing of the data elements for each of the ALS measures will be made available at a later date.

Testing and Implementation of the Measurement Set
The measures in the set are being made available without any prior testing. 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.

Desired Outcomes for Patients with ALS

Setting: Ambulatory and residential care (nursing facility, domiciliary, home care)



Purpose of Measurement Set

The American Academy of Neurology (AAN) formed an ALS Work Group to identify and define quality measures towards improving outcomes for patients with amyotrophic lateral sclerosis (ALS). The Work Group sought to develop measures to support the delivery of high quality care for patients with ALS. The Work Group developed measures that were focused on the gaps in care in need of significant improvement and the available rigorous clinical evidence for ALS. The Work Group considered the development of outcome, process, structural, composite, bundled, and group or system-level measures where it was appropriate.

Importance of Topic

Prevalence and Incidence

- ALS, also known as Lou Gehrig's disease is a type of motor neuron disease that is a rapidly progressive and fatal neurological disease.[1]
- 20-30,000 people in the US have ALS.[1]
- 5,000 people are diagnosed with ALS in the US annually.[1]
- ALS is one of the most common neuromuscular disease worldwide.[1]
- 90-95% of all ALS cases the disease occurs apparently at random with no clearly associated risk factors.[1]
- 5-10% of all ALS cases are inherited.[1]
- 20% of all familial cases result from a specific genetic defect that leads to mutation of the enzyme known as superoxide dismutase 1 (SOD1).[1]
- No cure exists for ALS. Newer pharmacotherapy agents have been found to reduce the progression, but not halt the disease development.[1]
- The prevalence of ALS is said to be between six and eight cases per 100,000 in the population. Using the higher prevalence estimate and data from the 2000 U.S. census, nearly 22,600 Americans are living with ALS at any one time. Since ALS is a disease of aging, as the U.S. population increases and ages, an increase in the prevalence of ALS can be anticipated.[2]
- Cognitive dysfunction is seen in 20–50%, while only 3–5% develop dementia that is usually of frontotemporal type.[3] Consensus criteria for diagnosis have recently been reported.[3]
- Death due to respiratory failure follows on average 2– 4 years after onset, but a small group may survive for a decade or more.[4]
- The mean age of onset is 47–52 years in familial cases (FALS) and 58–63 years in sporadic (SALS) cases.[5]
- The lifetime risk for developing ALS for individuals aged 18 years has been estimated to be 1 in 350 for men and 1 in 420 for women[5b] with male sex, increasing age and hereditary disposition being the main risk factors.[6]

Mortality and Morbidity

- Most patients with ALS die within 2 to 5 years of onset.[7] Only 10% of ALS patients survive for 10 years or more.[8]
- Treatment of respiratory insufficiency improves survival, quality of life and respiratory symptoms.[7,8] The diagnosis and management of respiratory insufficiency is critical because most deaths from ALS are due to respiratory failure.[7-10]
- Falls surveillance will lead to interventions to prevent falls and decrease fall related deaths in ALS patients. Falls are an independent predictor of adverse health outcomes.[11] Fall related deaths occur in 1.7% of ALS patients [12] Several specific risk factors for falls have been identified, including muscle weakness, deficits in gait or balance, visual deficits, arthritis, impairments in activities of daily living, depression, and cognitive impairment.[13]
- Studies confirm the presence of cognitive impairment in 50% of patients with ALS and particularly implicate executive dysfunction and mild memory decline in the disease process.[10] More severe impairment occurs in a subset of patients with ALS and has features consistent with frontal temporal dementia

(FTD).[14,15] Recent studies have demonstrated the feasibility of screening patients in a busy specialized ALS clinic,[16,17] but this is still not routinely practiced. A fuller characterization of the extent of cognitive and behavioral dysfunction in ALS has important implications given that it shortens survival [18], and the burden and stress for carers of patients with FTD is very great. It also has relevance to effective communication, legal issues and end-of-life decision making by patients with MND.[18]

- Pseudobulbar affect (PBA), excessive laughing or crying, or involuntary emotional expression disorder affects 20%–50% of patients with ALS, especially in pseudobulbar palsy.[19] Patients are embarrassed and isolated by these symptoms, which in turn greatly diminishes the patients' quality of life.
- Sialorrhea, or drooling, is embarrassing, socially isolating, and is associated with aspiration pneumonia. The prevalence is estimated at 50%, and 70% of patients receiving oral medications for treatment reported benefit (Class III).[10,20]
- Fatigue may be a symptom of depression, poor sleep, abnormal muscle activation, immobility, or respiratory dysfunction. Fatigue diminishes quality of life for patients with ALS. Fatigue was a side effect of therapy in 26% of patients taking riluzole vs. 13% taking placebo.[21] Asthenia occurred in 18% of patients taking riluzole vs. 12% of patients taking placebo in a larger study.[22]
- The prevalence of depression in ALS ranges from 0 to 44%, although systematic studies suggest 10% in advanced ALS (Class III).[10,24] Depression shortens survival and lowers quality of life for patients with ALS.[14] There is consensus among experts that depression should be treated in patients with ALS[10]; however, there are no controlled studies of benefit or harm.
- Insomnia is common in ALS and may be a symptom of early respiratory weakness, underlying anxiety, depression, or pain.[25] There is a concern that sedative/hypnotic agents may suppress the respiratory drive in patients with ALS.
- Weight loss is a key prognostic indicator for ALS with the risk of death increased 7-fold when body mass index is <18.5 kg/m2.[26-31]
- ALS patients have dysarthria in nearly all bulbar onset patients and nearly 40% of ALS patients with spinal onset. More than 95% of ALS patients cannot speak before death and patients who accept gastrostomy tube, non-invasive ventilation or tracheostomy-ventilation have a greater need for augmentative alternative communication as the disease progresses.[32-35]
- End of life discussions will improve patient decision making with respect to disease management.[1-6] Pain in ALS should be treated following accepted guidelines.[36-41]

Office Visits and Hospital Stays

- One study's significant findings were that common morbidities increased over time (pneumonia [38.1% to 47.3%], respiratory failure [26.9% to 35.5%], and nutritional deficiency [43.0% to 56.3%]); the median length of stay dropped from 6 to 4 days; mean hospital charges increased from $21,574 to $24,314; the proportion of hospital deaths decreased over time (17.6% to 14.6%), whereas the proportion discharged to home health/hospice care (14.0% to 18.2%) and to long-term care facilities (13.2% to 27.9%) increased. The odds ratio (OR) of death was 5.03 (95% CI: 4.57 to 5.54) for those admitted with respiratory failure, 1.36 (1.24 to 1.50) for those with pneumonia, and 0.84 (0.77 to 0.92) for those with nutritional deficiency. The high OR of death in patients admitted for pneumonia or respiratory failure is likely associated with more advanced disease, whereas the protective effect of admission for nutritional deficiency is consistent with the predominance of bulbar symptoms and admission earlier in the disease. The trends during the 15 years of this administrative data set were for increasing comorbidities and higher utilization of end-of-life care.[42]

Family Caregiving

- Caregiver burden was correlated to their level of depression and quality of life and, differently from other chronic disorders, increased with the worsening of patients' disability. ALS patients have a good objective perception of their impact on caregivers.[43]

- Recent studies assessing caregivers' burden in chronic neurologic disorders have found some features shared by caregivers: The perceived burden exceeds the objective measures of patients' impairment, the amount of burden is independent of diagnosis, and the patients' cognitive functioning is an important factor in determining the level of burden.[44]
- In one study seven ventilator-dependent ALS patients and eleven caregivers were interviewed in order to assess the impact of ventilator-dependence on patients and their families. The ALS Care Database questionnaires were administered with special attention to components derived from the Health Status Survey (SF-12) and ALS Quality-of-Life Index (ALSQLI) as well as the ALS Patient Caregiver Form. Six patients had difficulty communicating and one patient was totally unable to communicate. Patients had maximal limitation of daily activities as measured by The ALS QLI, yet a self-reported satisfactory quality-of-life. Caregivers were heavily burdened and their outside activities were severely limited. [45]
- In one study a total of 69 patient–caregiver pairs participated. For measures of physical function, _ ranged from 0.49 to 0.83, indicating moderate to excellent agreement. Patient and caregiver composite ALSFRS-R scores were highly correlated (r =0.92, p < 0.001). Agreement between patients and caregivers was high for ratings of patient pain, control over ALS, optimism, and will to live, and this level of agreement remained high over multiple assessments. In pairwise analyses, caregivers rated patients as having less energy, greater suffering, and greater weariness than patients indicated for themselves. Whereas patients rated caregivers as more burdened than caregivers reported for themselves. Caregivers can accurately report information about a patient's physical function at the end of life. However, patients and caregivers each overestimated the psychosocial impact of the disease on the other.[46]
- In one study thirty-one ALS patient– caregiver couples were interviewed at baseline and after 9 months. The mean ALS-FRS score was 28.7 (SD 7) at baseline and 24.1 (6.9) at the second interview (p=0.0001). Patients' mean MQoL score slightly increased from 6.8 (1.6) to 7 (1.1) (p=0.07); their ZDS score slightly increased (43.2 [8.7] at baseline and 45.7 [9.3] at the second interview) but they remained in the not depressed range. Caregivers' mean MQoL score slightly decreased, and their mean ZDS increased from 38.9 (8.1) to 42.2 (8.7) (p=0.02). The mean CBI score increased from 50.3 (17.6) to 55.8 (16.4) (p=0.03). They found a substantial steadiness of quality of life and depression in patients with amyotrophic lateral sclerosis over a 9-month period, vs. a significant increase of burden and depression of their caregivers. [47]

Cost

- Amyotrophic lateral sclerosis (ALS) is a difficult to diagnose, fatal, progressive degenerative disease with an average survival time of 2 to 5 years. Percutaneous endoscopic gastrostomy (PEG) and bi-level intermittent positive pressure (BIPAP) ventilation may be the major interventions leading to longer survival of patients with ALS. Riluzole has been shown to have modest effects on survival (as opposed to functional) gains and is currently the only drug approved for the treatment of ALS. Mechanical ventilation (via a tracheostomy tube) is expensive, but is widely used in later stage patients with ALS in the US. A review of nine cost-effectiveness studies of riluzole found the following: drug costs and survival gains are the major drivers of cost effectiveness; survival gains are estimated from truncated databases with a high degree of uncertainty; more accurate stage-specific utility weights based on patients who agreed to treatment are needed; case incidence-based evaluations should be carried out; cost-effectiveness ratios are insensitive to discount rates; employment and caregiver issues or externalities have been widely ignored; threshold acceptance cost-effectiveness values are ill-defined and evaluations are not generalizable to other countries because of cost and treatment style differences. On account of the high degree of uncertainty pertaining to survival gains and the relatively high costs per life years or quality-adjusted life-years gained, and while acknowledging that not every therapy has to be cost effective (e.g. orphan drugs), it is still inconclusive as to whether or not riluzole can be considered as cost-effective therapy for ALS.[48]
- The average cost for the power chairs was $26,404 (range, $19,376-$34,311), and the average cost a month is $917. Overall, 88% of respondents said they would get the same type of chair with the same features again, and 81% felt that the chair was a good value for the cost. We obtained first-hand knowledge from 32 patients with ALS/MND who are current PWC users on their use and satisfaction with their PWCs from

initial to current use. Based on this survey, patients with ALS/MND seen for their wheelchair evaluation with experienced clinicians exhibit high use and satisfaction with their PWCs.[49]

- This study reports the results of a long-term economic evaluation of riluzole in the treatment of amyotrophic lateral sclerosis (ALS) versus best supportive care in the United Kingdom. Applying the Markov model and extending the transitional probabilities using linear interpolation, the base case cost per life year gained was estimated at £15,192 while applying Standard Gamble utility scores, the base case cost per quality-adjusted life-year (QALY) was assessed at £22,086. Carrying out a probabilistic sensitivity analysis, the cost per QALY was estimated at £22,236 with standard deviation of £612. The results of the long-term analysis also show that riluzole on average increases survival in ALS patients by 6 months with approximately 5 months of the additional life gained in the early disease states, of which 4 months is spent in disease state 2, where quality of life is relatively high.[50]
- In one study a cost-utility analysis was used determine a priori what magnitude of health-related quality of life (HRQL) improvement early NIPPV initiation would need to achieve to be cost-effective in a future clinical trial. Using a Markov decision analytic model we calculated the benefit in health-state utility that NIPPV initiated at ALS diagnosis must achieve to be cost-effective. The primary outcome was the percent utility gained through NIPPV in relation to two common willingness-to-pay thresholds: 50,000 dollars and 100,000 dollars per quality-adjusted life year (QALY).Our results indicate that if NIPPV begun at the time of diagnosis improves ALS patient HRQL as little as 13.5%, it would be a cost-effective treatment. Tolerance of NIPPV (assuming a 20% improvement in HRQL) would only need to exceed 18% in our model for treatment to remain cost-effective using a conservative willingness-to-pay threshold of 50,000 dollars per QALY. If early use of NIPPV in ALS patients is shown to improve HRQL in future studies, it is likely to be a cost-effective treatment. Clinical trials of NIPPV begun at the time of ALS diagnosis are therefore warranted from a cost-effectiveness standpoint.[51]
- Improved survival was seen in patients with ALS attending tertiary ALS centres, independently from all other known prognostic factors, possibly through a better implementation of supportive treatments. Moreover, because of these centres, the hospitalisation rate was markedly reduced, thus offering a cost-effective service to patients with ALS and to the community as a whole.[52]
- In one study authors compared the cost of hospice care provided to 25 amyotrophic lateral sclerosis (ALS) patients and 159 lung cancer patients by the Wissahickon Hospice of the University of Pennsylvania. The mean length of stay was 86.7 days for ALS patients and 35.0 days for patients with lung cancer (P = .011). The mean per patient cost was 5622.93 dollars for the ALS patients and 2658.91 dollars for patients with lung cancer (P = .057) The average operating margin excluding administrative costs was 5293.04 dollars for ALS patients and 2126.74 dollars for patients with lung cancer (P = .008). The longer length of stay (LOS) accounts for this difference. Longer LOS can be accomplished by close clinical monitoring of ALS patients for the development of life threatening respiratory and/or nutritional compromise and by liberalizing the present hospice admission guidelines.[53]
- Neuromuscular disorders (NMD) are chronic devastating diseases. The aim of this multicenter cross-sectional study was to evaluate the socioeconomic impact of three NMDs in Germany. Patients (n = 107) with amyotrophic lateral sclerosis (ALS), myasthenia gravis (MG) or facioscapulohumeral muscular dystrophy (FSHD) were recruited consecutively in seven centers in Germany. The health-economic data were collected using a "bottom-up" approach consisting of comprehensive questionnaires and patient diaries. Costs were evaluated from the societal perspective in 2009 Euros (EUR). Total annual costs from the societal perspective were EUR 36,380 (95% CI 27,090-47,970) per patient in ALS, EUR 26,240 (95% CI 17,770-37,940) in FSHD and EUR 14,950 (95% CI 10,470-21,730) in MG. The main components of costs were the expenditures of health insurance and the loss of productivity of patients and their caregivers. The following independent cost-driving factors were identified: disease severity, assistance in activities of daily living (ADL), dementia and younger age in ALS, disease severity in FSHD and assistance in ADL, disease severity and assistance in ADL in MG.[54]
- The gap in current research should not be interpreted as proof that multidisciplinary care is ineffective. Further research into types of appropriate studies, caregiver needs and various aspects of multidisciplinary care in the MND population is needed.[55]

Opportunity for Improvement

- Treatments for ALS are underutilized even in specialized clinics.[56,57] Studies suggest that even in tertiary care centers, there are varying degrees of adherence to the evidence-based AAN Practice parameters.[1,2] Recent studies show that there is a much higher utilization rate of evidence-based treatments in multidisciplinary clinics than in community-based care.[57]Data are especially indicative of underuse of riluzole (60% of patients), PEG (9%), and noninvasive ventilation (22%), with greatest gains in utilization occurring in the specialized ALS clinics. These important treatments lengthen life and improve quality of life, but they are neglected by many patients and health care professionals.[56,57] Moreover, the level of satisfaction with the rendering of the diagnosis and overall satisfaction with care is significantly higher for patients attending a multidisciplinary clinic.[57]
- Utilization of therapies increased from 1997 before publication of the ALS Practice parameters from 3-31%, up to 9-43% in 2004.[57] Details below, documenting persistent underutilization of many therapies, especially PEG (16%) and NIV (9%).The most recent analysis of 5,600 patients shows interesting epidemiological observations and treatment trends. Proper management of many ALS symptoms has increased substantially since the first publication of the guidelines, and awareness of pseudobulbar affect has increased. Other recommendations are underutilized: Only 9% undergo percutaneous endoscopic gastrostomy, although this procedure was recommended in 22% of patients; and noninvasive positive pressure ventilation was used by only 21% of patients despite being associated with improved 5-year survival rates.[57] The following data indicate the increase in utilization of therapies for ALS between 1999 and 2004. All are significant increases ($p<0.0001$)
  Patients taking riluzole (%) 45 to 56.
  Patients taking medication for sialorrhea (%) 12 to 17.
  Patients receiving treatment for pseudobulbar affect (%) 28 to 42 .
  Patients receiving treatment for depression (%) 23 to 42
  Patients receiving treatment for sleep disturbance (%) 17to 29
  Patients receiving PEG when FVC was 50% of predicted (%) 9 to 16
  Patients receiving NIPPV when FVC was 40% of predicted (%) 5 to 9
  Patients taking nutritional supplements (%) 9 to 19
  Patients receiving physical therapy (%) 31 to 43
  Patients receiving occupational therapy (%) 19 to 31
- Riluzole is currently the only available disease modifying pharmacotherapy available to slow down progression of ALS. Only 60% of patients are taking the riluzole in the United States, compared to nearly 100% in European countries (France, Italy, Germany).[57] This utilization is improved compared to 45% in 1997, a rise that reflects increased awareness and experience of treating physicians.[56] These data reflect the utilization of riluzole in large multidisciplinary clinics, and it is much lower in community-treated patients. Considerable misunderstanding exists around safety and efficacy, both for patients and physicians. More education is needed. The most influential factor in whether patients take riluzole is the knowledge and enthusiasm of the treating physician (Bryant et al). ALS experts in a multidisciplinary clinic are most likely to adequately inform patients about this neuroprotective medication. Also, the more recent registry studies suggesting a much greater survival benefit have been impressive.[58]
- There is now considerable evidence for cognitive and behavioral manifestations in ALS. The domain of cognitive and behavioral impairment in ALS is a rapidly evolving field and there is little consensus regarding diagnostic criteria and assessment methods.[59] Estimates of cognitive impairment range from 10%[60] to 75% [61] in those diagnosed with ALS. A population-based sample produced an estimate of 28%.[62] The prevalence of impairment meeting criteria for dementia ranged from 15%[63] to 41%.[64] Behavioral impairment (irritability and social disinhibition) was identified in 39%.[65] Although there has not been a systematic study of how many clinics do screening, there is good support for the presence of a gap.[66] A recent large study found that most patients and caregivers were not informed about the presence of cognitive or behavioral/ psychological impairment. Patients commonly reported being told by their doctor about physical symptoms such as problems walking (85%) or stiffness/cramps (74%) but not psychological

issues like emotional liability (46%) or cognitive change (11%). Patients and caregivers have indicated that they do want to know about whether they are so affected. These data suggest that screening is not being done widely at all.[66]

- Treatment for both refractory sialorrhea and pseudobulbar affect are underutilized in ALS.[56,57] Many practitioners are unaware of the condition and also of the recently approved treatment for pseudobulbar affect.[67] Most other symptoms of ALS are treatable, albeit with less evidence base. Still, most symptoms in ALS can be treated and studies suggest that these treatments are underutilized.[56,57]
- The vast majority of patients with ALS only get standard vital capacity measures to monitor pulmonary function, and in many instances even this basic measurement is not made. The use of nocturnal oximetry, polysomnography, sniff nasal pressure (SNP), maximum inspiratory pressure (MIP), peak expiratory cough flow (PCEF) and supine vital capacity (VC) should permit earlier detection and treatment of respiratory insufficiency in ALS.[7,9,10] More studies are needed to clarify which of these measures is the best, but several studies have shown that MIP is probably the most sensitive indicator of early diaphragm weakness.[7,68,69] Symptoms of early respiratory insufficiency should be specifically asked for: dyspnea, orthopnea, excessive daytime sleepiness, insomnia, fatigue, morning headache. Patients often do not volunteer these symptoms and the clinician may detect early respiratory insufficiency by enquiring about them, which is often not done.[3,10,57]
- Survival is improved with increased proportion of use of non-invasive ventilation from 16% of ALS patient with respiratory insufficiency to 51%[71,72], while over 65% of ALS patients have said that they would want respiratory support indicating that noninvasive ventilation is still underutilized in ALS patients.[56,57, 72] Even though published guidelines to date have made a positive impact in doubling utilization rates, underutilization of this important treatment is still evident.[56,57]
- Prevalence of malnutrition varies between 16 - 55% in ALS patients across several studies.[73-75]
- Not all patients who should have enteral feeding receive this treatment with only 43% of ALS patients who met guideline indications for gastrostomy tube placement undergoing the procedure. [57] Increasing proportions of patients who should receive this treatment are now receiving treatment but not all. [56, 72,76,77]
- Speech assessment in ALS patients identifies dysarthria, independent of dysphagia that limits communication and maintenance of their active communicator role. Speech correction should focus on the maintenance of functional communication. Assessment should involve measurement of speech rate (words per minute). Augmentative communication is recommended when speech rate is less than 125 words per minute. Nearly 88% of ALS patients are evaluated by this criterion, but fewer than half implement appropriate interventions. [78-82]
- End of life treatment for ALS patients is an evolving practice. Between 60-88% of patients die in a medical facility in some countries and not at home, while over 58% of seriously ill ALS patients do not have hospice care.[12,83,84] Many patients are not adequately informed about advance directives and end of life decision making and many hospice workers are not familiar with ALS. [56,57,85] Changes in the protocols for end of life care vary widely and over time.[53,84]
- Patients with neurologic or general conditions associated with an increased risk of falling should be asked about recent falls and further examined for the presence of specific neurologic deficits that predict falls, which include gait and balance disorders; deficits of lower extremity strength, sensation, and coordination; and cognitive impairments. If substantial risks of falls are identified, appropriate interventions that are described in other evidence-based guidelines may be considered.[86] There is an increased rate [Hazard ratio = 6.5 (95% CI 2.4-14.1)]of falls with head injury in ALS patients compared with a control population in the first year of ALS.[87] There is an increased prevalence of falls in ALS patients of 0.7 falls per patient per 6 months.[88]

Disparities

- All races and ethnic backgrounds are affected by ALS.[1]
- ALS most common in individuals 40-60 years old, but younger and older people can develop the disease.[1]

- Men are more likely to develop ALS than women. Studies suggest an overall ratio of about 1.5 men to every woman who develops ALS in Western countries.[2]

Clinical Evidence Base

Clinical practice guidelines serve as the foundation for the development of performance measures. Relatively few clinical practice guidelines have been developed for ALS; however there are a number of reputable consensus papers, systematic reviews and Cochrane reviews which offered a robust evidence base to guide clinical decision-making and performance measure development. Guidelines from the American Academy of Neurology,[89-91] European Federation of Neurological Sciences,[92,93] British Thoracic Society/Association of Chartered Physiotherapists in Respiratory Care,[94] National Institute for Health and Clinical,[95] two systematic reviews,[96,97] and one Cochrane review.[98]

Relevant guidelines met all of the required elements and many, if not all, of the preferred elements outlined in a PCPI position statement establishing a framework for consistent and objective selection of clinical practice guidelines from which PCPI Work Groups may derive clinical performance measures.[99,100]

Performance measures, however, are not clinical practice guidelines and cannot capture the full spectrum of care for all patients with ALS. The guideline principles with the strongest recommendations and often the highest level of evidence (well-designed randomized-controlled trials) served as the basis for measures in this set.

ALS Outcomes

Ideally, a set of performance measures would include both measures of outcomes as well as measures of processes that are known to positively influence desirable outcomes. The development of outcome measures for ALS proved particularly challenging given the progressive nature of the disease. The goals are often focused on improving the quality of life for patients, maintaining optimal function and providing maximum comfort. The Work Group sought to identify the desired outcomes for ALS with a goal of developing performance measures based on processes that are associated with desired outcomes and reflect high quality care.

Desired outcomes for ALS include:

1. Care coordination between multiple providers
2. Enhance patient quality of life
3. Slowing ALS disease progression
4. Minimize ALS symptoms
5. Educate patients and promote patient-centered decision making
6. Avoid emergent decision making/promote advanced planning
7. Reduce hospitalizations

Intended Audience, Care Setting, and Patient Population

The AAN encourages use of these measures by physicians and other health care professionals, where appropriate, to manage the care for all patients with amyotrophic lateral sclerosis.

ALS Work Group Recommendations

The measurement set includes measures that focus on accurate and appropriate evaluation and monitoring of disease status and associated symptoms to guide treatment, therapeutic options in eligible patients, increasing patient awareness of advanced planning, and patient safety. The ALS Work Group identified several desired outcomes for patients with ALS (see "Link to Outcomes" diagram in preceding section). Current quality gaps in ALS care emphasize the need to improve specific processes that have been demonstrated to improve ALS outcomes. As a result, many of the measures in the ALS set focus on the provision of effective and patient-centered care. These clinical performance measures are designed for practitioner level quality improvement to achieve better outcomes for patients with ALS. Unless otherwise indicated, the measures are also appropriate for accountability if the appropriate methodological, statistical, and implementation rules are achieved.

Amyotrophic Lateral Sclerosis Measures
Measures addressing accurate and appropriate evaluation/monitoring of disease status and associated symptoms to guide treatment options
Measure #1: ALS Multidisciplinary Care Plan Developed or Updated
Measure #3: ALS Cognitive and Behavioral Impairment Screening
Measure #4: ALS Symptomatic Therapy Treatment Offered
Measure #5: ALS Respiratory Insufficiency Querying and Referral for Pulmonary Function Testing
Measure #7: ALS Screening for Dysphagia, Weight Loss and Impaired Nutrition
Measure #8: ALS Nutritional Support Offered
Measure #9: ALS Communication Support Referral
Measures addressing effective therapeutic options in eligible patients
Measure #2: Disease Modifying Pharmacotherapy for ALS Discussed
Measure #6: ALS Noninvasive Ventilation Treatment for Respiratory Insufficiency Discussed
Measures addressing increasing patient awareness of advanced planning
Measure #10: ALS End of Life Planning Assistance
Measures addressing patient safety
Measure #11: ALS Falls Querying*

Amyotrophic Lateral Sclerosis Measures

*This measure is for quality initiatives only.

Institute of Medicine Domains of Health Care Quality

Measure	Safe	Effective		Patient-Centered	Timely	Efficient	Equitable
		Underuse	Overuse				
Measure #1: ALS Multidisciplinary Care Plan Developed or Updated		X					
Measure #2: Disease Modifying Pharmacotherapy Discussed	X	X			X		
Measure #3: ALS Cognitive and Behavioral Impairment Screening		X					
Measure #4: ALS Symptomatic Therapy Treatment Offered		X			X		
Measure #5: ALS Respiratory Insufficiency Querying and Referral for Pulmonary Function Testing		X					
Measure #6: ALS Noninvasive Ventilation Treatment for Respiratory Insufficiency Discussed		X					
Measure #7: ALS Screening for Dysphagia, Weight Loss and Impaired Nutrition		X					
Measure #8: ALS Nutritional Support Offered		X		X	X		
Measure #9: ALS Communication Support Referral		X		X			

Measure #10: ALS End of Life Planning Assistance		X		X	X		
Measure #11: ALS Falls Querying*	X	X		X			

Other Potential Measures

The Work Group considered several other important constructs in ALS care, though ultimately determined that they were not appropriate as the subject of performance measures. The Establishing the diagnosis, and the delivery of the diagnosis, of ALS was felt to be very important by the work group. However, there was not enough evidence to support the development of a measure. In addition, several individual measures for symptoms of ALS were initially drafted by the work group. However, the decision was made to combine the symptoms into one measure as the work group felt it was more important to not overburden the individual practitioner with multiple measures about ALS symptoms. The work group also considered creating an ALS patient experience of care measure. However, there was insufficient evidence to create a measure and the work group felt that there may be current survey and screening tools already validated that could be applied to patients with ALS.

Measure Harmonization

When existing hospital-level or plan-level measures are available for the same measurement topics, the AAN attempts to harmonize the measures to the extent feasible. The AAN works to ensure there is no duplication of existing measures. The AAN reaches out to partner organizations for input on the measures, involves key stakeholders on the measure development workgroups and posts the measures during a 30 day public comment period for comment by any interested individual or group. There remains a scarcity of measures to address the quality of ALS care.

There exist two other measures that refer to advanced care planning. The American Geriatrics Society (2008) has a measure "Percentage of patients aged 65 years and older who have an advance care plan or surrogate decision maker documented in the medical record or documentation in the medical record that an advance care plan was discussed but the patient did not wish or was not able to name a surrogate decision maker or provide an advance care plan." However this measure is limited to those over the age of 65. The work group felt it was important to not exclude patients under 65 years old who have ALS from an end of life planning measure. In addition, the Institute for Clinical Systems Improvement (ICSI) (2009) has a measure that states the "Percentage of adult patients with the specified progressive, debilitating disease who have a palliative care plan* in chart." *A completed palliative care plan addresses all seven domains of care: physical aspects, cultural aspects, psychological aspects, social aspects, spiritual/ religious/existential aspects, ethical/legal aspects, and care of the imminently dying patient. However, this measure does not reference specific end of life needs that are relevant for patients with ALS. This measure was also not developed by a medical specialty society and the methods used to develop the measure are unclear.

For the 2012 Physician Quality Reporting System (PQRS) program, there are three measures that relate to falls. PQRS #154 is a Falls Risk Assessment, #155 is about a plan of care, and #318 is screening for future fall risk. However, all three refer to only patients 65 years and older, and many ALS patients will not fit the denominator, so therefore, a measure specific to ALS would not be overlapping.

Technical Specifications Overview

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

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

Because administrative claims are currently the only available sources of data, specifications to collect and report on the Amyotrophic Lateral Sclerosis measures for administrative claims are included in this document.

The AAN is in the process of creating data elements required for electronic capture with Electronic Health Records (EHRs). A listing of the data elements for each of the ALS measures will be made available at a later date.

Measure Exclusions

For *process measures*, the AAN follows the PCPI's three categories of reasons for which a patient may be excluded from the denominator of an individual measure:

- Medical exclusion examples:

- not indicated (absence of organ/limb, already received/performed, other)
- contraindicated (patient allergic history, potential adverse drug interaction, other)

- Patient exclusion examples:

- patient declined
- social or religious reasons
- other patient reasons

- System exclusion examples:

- resources to perform the services not available
- insurance coverage/payer-related limitations
- other reasons attributable to health care delivery system

These measure exclusion categories are not available uniformly across all measures; for each measure, there must be a clear rationale to permit an exclusion for a medical, patient, or system reason. For some measures, examples have been provided in the measure exclusion language of instances that would constitute an exclusion. Examples are intended to guide clinicians and are not all-inclusive lists of all possible reasons why a patient could be excluded from a measure. The exclusion of a patient may be reported by appending the appropriate modifier to the CPT Category II code designated for the measure:

- Medical reasons: modifier 1P
- Patient reasons: modifier 2P
- System reasons: modifier 3P

Although this methodology does not require the external reporting of more detailed exclusion data, the AAN follows the PCPI's recommendation that physicians document the *specific* reasons for exclusion in patients' medical records for purposes of optimal patient management and audit-readiness.[98] The PCPI also advocates the systematic review and analysis of each physician's exclusions data to identify practice patterns and opportunities for quality improvement. For example, it is possible for implementers to calculate the percentage of patients that physicians have identified as meeting the criteria for exclusion.

Please refer to documentation for each individual measure for information on the acceptable exclusion categories and the codes and modifiers to be used for reporting.

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 ALS 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.

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Measure #1: ALS Multidisciplinary Care Plan Developed or Updated

Amyotrophic Lateral Sclerosis

Measure Description

Percentage of patients diagnosed with ALS for whom a multi-disciplinary care plan* was developed, if not done previously, and the plan was updated at least once annually.

*Multi-disciplinary care plan should include a neurologist and at least four of the following specialists: pulmonologist, gastroenterologist, physiatrist, psychiatrist, social worker, occupational therapist, physical therapist, speech language pathologist, psychologist, respiratory therapist, genetic counselor, palliative care specialist, specialized nurse, dietician, or dentist.

Measure Components

Numerator Statement	Patients for whom a multi-disciplinary care plan* was developed, if not done previously, and the plan was updated at least once annually. *Multi-disciplinary care plan should include a neurologist and at least four of the following specialists: pulmonologist, gastroenterologist, physiatrist, psychiatrist, social worker, occupational therapist, physical therapist, speech language pathologist, psychologist, respiratory therapist, genetic counselor, palliative care specialist, specialized nurse, dietician, or dentist.
Denominator Statement	All patients with a diagnosis of amyotrophic lateral sclerosis.
Denominator Exclusions	**The following clinical recommendation statements are quoted verbatim from the referenced clinical guidelines and represent the evidence base for the measure:** • Documentation of a system reason for not developing and updating annually a multi-disciplinary care plan (eg patient has no insurance to cover a multidisciplinary plan)
Supporting Guideline & Other References	• Specialized multidisciplinary clinical referral should be considered for management of patients with ALS to optimize health care delivery (Level B) and prolong survival (Level B) and may be considered to enhance quality of life (Level C).[1] • Multidisciplinary care should be available for people affected by ALS as attendance at a multidisciplinary clinic improves care, and may extend survival.[2] (GPP) • The following specialties should be part of or be readily available to the multidisciplinary team: a consultant in neurology, pulmonologist, gastroenterologist, rehabilitation medicine physician, social counselor, occupational therapist, speech language pathologist, specialized nurse, physical therapist, dietitian, psychologist, dentist.[2](GPP) • Initiate discussions about all treatment options such as non-invasive, invasive ventilation and terminal phase treatment as soon as symptoms or signs of respiratory problems develop. Discussions should be as early as possible to enable advance planning or directives (Level II; note this is a systematic review).[3] • A palliative care approach should be incorporated into the care plan for patients and carers from the time of diagnosis (Class III recommendation; note this is a systematic review).[4] [1]Miller RG, Jackson CE, Kasarskis EJ, et al. Practice Parameter update: The care of the patient with amyotrophic lateral sclerosis: Multidisciplinary care, symptom management, and cognitive/behavioral

	impairment (an evidence-based review): Report of the Quality Standards Subcommittee of the American Academy of Neurology *Neurology* 2009:73:1227-1233 [2]Andersen PM, Borasio GD, Dengler R, et al. EFNS task force on management of amyotrophic lateral sclerosis: guidelines for diagnosing and clinical care of patients and relatives. *European J of Neurology* 2005;12:921-938 (GPP=Good Practice Point) [3] Heffernan C, Jenkinson C, Holmes T, et al. C. Management of respiration in MND/ALS patients: An evidence based review. *Amyotrophic Lateral Sclerosis* 2006; 7(1):5-15. (Systematic Review) [4] Tripodoro VA, De Vito EL. Management of dyspnea in advanced motor neuron diseases. *Curr Opin Support Palliat Care* 2008; 2(3):173-9. (Systematic Review)

Measure Importance

Relationship to desired outcome

In specialized multidisciplinary clinics, patients with ALS receive comprehensive care from a neurologist, pulmonologist, gastroenterologist, physiatrist, social worker, occupational therapist, speech language pathologist, respiratory therapist, specialized nurse case manager, physical therapist, dietitian, psychologist, dentist, and/or palliative care expert.[1,2] Moreover, the level of satisfaction with the rendering of the diagnosis and overall satisfaction with care is significantly higher for patients attending a multidisciplinary clinic.[2]

Specialized clinics coordinate care and interface with a primary care physician, local neurologist and community-based services. Patients who attend specialized ALS clinics are younger and have longer symptom duration than neurology clinic patients, indicating possible referral bias.[3] Patient care and survival were examined for 97 patients attending specialized ALS clinics in Italy compared with 124 patients in neurology clinics.[4] There was increased utilization of riluzole, percutaneous endoscopic gastrostomy (PEG), and noninvasive ventilation (NIV) in the ALS clinics, and fewer hospital admissions. Mean survival was longer in specialized ALS clinics (1,080 days vs. 775 days, p=0.008). Using COX multivariate analysis, attending an ALS specialized clinic independently predicted longer survival for patients.

Prolonged survival (7.5 months, p<0.0001) was found for patients in Ireland attending multidisciplinary ALS clinics.[5] Patients at ALS clinics were younger and more likely to receive riluzole (99% vs. 61%). Multidisciplinary care was an independent predictor of survival (p=0.02) and reduced the risk of death by 47% in a 5-year study.[5] Dutch patients in multidisciplinary ALS clinics (n=133) were compared with 75 patients receiving general care[6] (6). Patients were well-matched and data were collected by a blinded nurse. Patients in multidisciplinary clinic received more aids and appliances (93% vs. 81%, p =0.008) and had higher quality of life (SF-36® Health Survey, p <0.01). Beneficial effects derived from a single visit to a multidisciplinary clinic, suggesting better coordination of care. Importantly, patients attending multidisciplinary clinics had fewer hospital admissions and shorter inpatient stays than those cared for in the community.

By contrast, another study, in Southern Italy, documented no increase in survival from attendance at a multidisciplinary clinic.[7] Riluzole use was higher in patients attending a multidisciplinary clinic (61% vs. 43%, p = 0.02) but very few patients received PEG (6% vs. 2%) or NIV (2% in each group). There was a non-significant 10% increase in survival in those attending a multidisciplinary clinic after 12 months. Low utilization of palliative care, case management, PEG, NIV, and riluzole, compared with the 3 positive studies above, may account for the lack of survival benefit in this study.

Thus, three studies show that multidisciplinary clinics specializing in ALS care are probably effective in several ways: increased use of adaptive equipment; increased utilization of riluzole, PEG, and NIV; improved quality of life; and lengthened survival. However, one study with

	low use of these treatments found no survival benefit. **References** [1] The EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis:, Andersen PM, Abrahams S, Borasio GD, et al. EFNS guidelines on the Clinical Management of Amyotrophic Lateral Sclerosis (MALS) - revised report of an EFNS task force. *Eur J Neurol* 2011 Sep 14. [Epub ahead of print] [2] Miller RG, Jackson CE, Kasarskis EJ, et al. Quality Standards Subcommittee of the American Academy of Neurology. Practice parameter update: The care of the patient with amyotrophic lateral sclerosis: multidisciplinary care, symptom management, and cognitive/behavioral impairment (an evidence-based review): report of the Quality Standards Subcommittee of the American Academy of Neurology. *Neurology* 2009; 73(15):1227-33. [3] Sorenson EJ, Mandrekar J, Crum B, Stevens JC. Effect of referral bias on assessing survival in ALS. *Neurology* 2007; 68:600-2. [4] Chio A, Bottacchi E, Buffa C, Mutani R, Mora G. Positive effects of tertiary centres for amyotrophic lateral sclerosis on outcome and use of hospital facilities. *J Neurol Neurosurg Psychiatry* 2006; 77:948-50. [5] Traynor BJ, Alexander M, Corr B, et al. Effects of a multidisciplinary ALS clinic on survival. *J Neurol Neurosurg Psychiatry* 2003;74:1258-61. [6] Van den Berg JP, Kalmijn S, Lindeman E, et al. Multidisciplinary ALS care improves quality of life in patients with ALS. *Neurology* 2005;65:1264-67. [7] Zoccolella S, Beghi E, Palagano G, et al. ALS multidisciplinary clinic and survival : Results from a population-based study in Southern Italy. *J Neurol* 2007; 254:1107–1112.
Opportunity for Improvement	Treatments for ALS are underutilized even in specialized clinics.[1,2] Studies suggest that even in tertiary care centers, there are varying degrees of adherence to the evidence-based AAN Practice parameters.[1,2] Recent studies show that there is a much higher utilization rate of evidence-based treatments in multidisciplinary clinics than in community-based care.[2] Data are especially indicative of underuse of riluzole (60% of patients), PEG (9%), and noninvasive ventilation (22%), with greatest gains in utilization occurring in the specialized ALS clinics. These important treatments lengthen life and improve quality of life, but they are neglected by many patients and health care professionals.[1,2] Access to the limited number of ALS specialized clinics may involve long distance travel which may be a barrier for patients who are unable to travel to an ALS clinic. Telemedicine might be a solution to this challenge. **References** [1]Bradley WG, Anderson F, Gowda N, Miller RG. Changes in the management of ALS since the publication of the AAN ALS practice parameter 1999. *Amyotroph Lateral Scler Other Motor Neuron Disord* 2004; 5:240-44. [2]Miller RG, Anderson F, Brooks BR, set al. ALS CARE Study Group. Outcomes research in amyotrophic lateral sclerosis: lessons learned from the amyotrophic lateral sclerosis clinical assessment, research, and education database. *Ann Neurol* 2009; 65(1):S24-8.
IOM Domains of Health Care Quality Addressed	Effective Patient Centered
Exclusion Justification	A system reason exclusion has been included for patients who have no insurance to cover the cost of a multidisciplinary care plan.
Harmonization with Existing Measures	There are no other measures currently available that are similar to this measure or need to be harmonized with this measure.

Measure Designation

Measure purpose	• Quality improvement • Accountability
Type of measure	• Process
Level of Measurement	• Individual practitioner
Care setting	• Ambulatory care setting
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

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 –CM Diagnosis Codes: 335.20 (amyotrophic lateral sclerosis) AND CPT E/M Service Code: 99201, 99202, 99203, 99204, 99205 (office-new patient), 99211,99212, 99213, 99214, 99215 (office-established patient), 99241, 99242, 99243, 99244, 99245 (outpatient consult), 99304, 99305, 99306, 99307, 99308, 99309, 99310 (nursing facility), 99324, 99325, 99326, 99327, 99328, 99334, 99335, 99336, 99337 (domiciliary), 99341, 99342, 99343, 99344, 99345, 99347, 99348, 99349, 99350 (home visit)
Numerator	Patients for whom a multi-disciplinary care plan* was developed, if not done previously, and the plan was updated at least once annually. *Multi-disciplinary care plan should include a neurologist and at least <u>four</u> of the following specialists: pulmonologist, gastroenterologist, physiatrist, psychiatrist, social worker, occupational therapist, physical therapist, speech language pathologist, psychologist, respiratory therapist, genetic counselor, palliative care specialist, specialized nurse, dietician, or dentist. Reporting Instructions: • For all patients meeting the denominator criteria, report the CPT Category II, **0580F**, *ALS multidisciplinary care plan developed or updated* 0580F *ALS multidisciplinary care plan developed or updated*

Denominator Exclusions	All patients with a diagnosis of amyotrophic lateral sclerosis. Reporting Instructions: • Documentation of a system reason for not developing and updating annually a multi-disciplinary care plan (eg patient has no insurance to cover rehabilitation or treatment plan) Reporting Instructions: o For patient with appropriate exclusion criteria, report **0580F-3P**

Measure #2: Disease Modifying Pharmacotherapy for ALS Discussed

Amyotrophic Lateral Sclerosis

Measure Description

Percentage of patients with a diagnosis of amyotrophic lateral sclerosis with whom the clinician discussed disease-modifying pharmacotherapy (riluzole) to slow ALS disease progression at least once annually.

Measure Components

Numerator Statement	Patients with whom the clinician discussed disease-modifying pharmacotherapy (riluzole**)** to slow ALS disease progression at least once annually.
Denominator Statement	All patients with a diagnosis of amyotrophic lateral sclerosis.
Denominator Exclusions	• No exclusions applicable for this measure.
Supporting Guideline & Other References	**The following clinical recommendation statements are quoted verbatim from the referenced clinical guidelines and represent the evidence base for the measure:** • Riluzole should be offered to slow disease progression in patients with ALS (Level A).[1] • Riluzole 50 mg twice a day is reasonably safe and probably prolongs median survival by about two to three months in patients with amyotrophic lateral sclerosis. (No level of evidence listed.) This is a Cochrane review.[2] • ALS patients should be offered treatment with riluzole 50 mg twice daily (Class 1A, GPP)[3] • Patients treated with riluzole should be monitored regularly for safety (Class 1A, GPP).[3] • Treatment should be initiated as early as possible after the patient has been informed of the diagnosis taking into account expected therapeutic benefits and potential safety issues (class 1A). Realistic expectations for treatment effects and potential side effects should be discussed with the patient and caregivers. (GPP)[3] • Treatment with riluzole should be considered in PMA and PLS patients who have a first degree relative with ALS. (GPP)[3] [1]Miller RG, Jackson CE, Kasarskis EJ, et al. Practice Parameter Update: The care of the patient with amyotrophic lateral sclerosis: Drug, nutritional, and respiratory therapies (an evidence-based review): Report of the Quality Standards Subcommittee of the American Academy of Neurology *Neurology* 2009;73:1218-1226 [2]Miller RG, Mitchell JD, Lyon M, More DH. Riluzole for amyotrophic lateral sclerosis (ALS)/motor neuron disease (MND) (Review). 2009 The Cochrane Library. 2:1-27. [3]Andersen PM, Borasio GD, Dengler R, et al. EFNS task force on management of amyotrophic lateral sclerosis: guidelines for diagnosing and clinical care of patients and relatives. *European J of Neurology* 2005;12:921-938 (GPP=Good Practice Point)

Measure Importance

Relationship to	Riluzole is approved by the Food and Drug Administration (FDA) for slowing disease

desired outcome

progress in ALS, and it is the only currently available disease modifying pharmacotherapy for ALS. Riluzole was the subject of a practice advisory published by the American Academy of Neurology in 1997.[1] The practice advisory recommended riluzole 50 mg BID to prolong survival for those with definite or probable ALS less than 5 years duration, with forced vital capacity (FVC) >60%, and without tracheostomy (Level A). Expert opinion suggested potential benefit for those with suspected or possible ALS with symptoms longer than 5 years, FVC >60%, and tracheostomy for prevention of aspiration only. Since 1997, 2 other controlled clinical trials have been published (Class I)[2,3] and all of the available evidence has been reviewed.[4] Riluzole has a modest beneficial effect in slowing disease progression (prolonged survival of 2–3 months) based on 4 Class I trials. The number needed to treat to delay 1 death until after 12 months was 11. However, 5 studies using large databases spanning 5 to 10 years have suggested that treatment with riluzole might be associated with a prolonged survival of 6 months (Class II)[5], 10 months (Class III)[6], 12 months (Class III)[7], 14 months (Class III)[8], or even 21 months (Class III)[9]. These cohort studies had longer-term follow-up than the clinical trials, but are subject to greater bias. After 10 years of patient experience, the drug appears to be safe but expensive. In fact, the cost does limit access to the drug for a significant portion of patients.[4,10] Fatigue and nausea are known side effects. Riluzole is safe and effective for slowing disease progression to a modest degree in ALS (4 Class I studies).

References

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Opportunity for Improvement

Riluzole is currently the only available disease modifying pharmacotherapy available to slow down progression of ALS. Only 60% of patients are taking the riluzole in the United States, compared to nearly 100% in European countries (France, Italy, Germany).[1] This utilization is improved compared to 45% in 1997, a rise that reflects increased awareness and experience of treating physicians.[2] The cost is still a major factor for many patients. These data reflect the utilization of riluzole in large multidisciplinary clinics, and it is much lower in community-treated patients. Considerable misunderstanding exists around safety and efficacy, both for patients and physicians. More education is needed. The most influential factor in whether patients take riluzole is the knowledge and enthusiasm of the treating physician.[3,4] ALS experts in a multidisciplinary clinic are most likely to adequately inform patients about this

neuroprotective medication. Also, the more recent registry studies suggesting a much greater survival benefit have been impressive.[3]

References

[1] Miller RG, Anderson F, Brooks BR, Mitsumoto H, Bradley WG, Ringel SP; ALS CARE Study Group. Outcomes research in amyotrophic lateral sclerosis: lessons learned from the amyotrophic lateral sclerosis clinical assessment, research, and education database. *Ann Neurol* 2009; 65 Suppl 1:S24-8.
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IOM Domains of Health Care Quality Addressed	Safe Effective Patient centered Timely
Exclusion Justification	No exclusions relevant for this measure.
Harmonization with Existing Measures	There are no other measures currently available that are similar to this measure or need to be harmonized with this measure.

Measure Designation

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

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 –CM Diagnosis Codes: 335.20 (amyotrophic lateral sclerosis) AND

	CPT E/M Service Code: 99201, 99202, 99203, 99204, 99205 (office-new patient), 99211,99212, 99213, 99214, 99215 (office-established patient), 99241, 99242, 99243, 99244, 99245 (outpatient consult), 99304, 99305, 99306, 99307, 99308, 99309, 99310 (nursing facility), 99324, 99325, 99326, 99327, 99328, 99334, 99335, 99336, 99337 (domiciliary), 99341, 99342, 99343, 99344, 99345, 99347, 99348, 99349, 99350 (home visit)
Numerator	Patients with whom the clinician discussed disease-modifying pharmacotherapy (riluzole**)** to slow ALS disease progression and the plan was updated at least once annually. Reporting Instructions: • For all patients meeting the denominator criteria, report the CPT Category II, **4540F**, *Disease modifying pharmacotherapy discussed* 4540F1 *Disease modifying pharmacotherapy discussed*
Denominator Exclusions	All patients with a diagnosis of amyotrophic lateral sclerosis. Reporting Instructions: • There are no exclusions for this measure. Do not report modifiers 1P, 2P or 3P with 4540F.

Measure #3: ALS Cognitive Impairment and Behavioral Impairment Screening

Amyotrophic Lateral Sclerosis

Measure Description

Percentage of patients diagnosed with ALS who are screened at least once annually for cognitive impairment (eg frontotemporal dementia screening or ALS Cognitive Behavioral Screen (CBS)) and behavioral impairment (eg ALS CBS).

Measure Components

Numerator Statement	Patients who are screened at least once annually for cognitive impairment (eg frontotemporal dementia screening or ALS Cognitive Behavioral Screen (CBS)) and behavioral impairment (eg ALS CBS).
Denominator Statement	All patients with a diagnosis of amyotrophic lateral sclerosis.
Denominator Exclusions	• Documentation of a medical reason for not screening the patient for cognitive and behavioral impairment (eg patient currently diagnosed with severe cognitive impairment) • Documentation of a patient reason for not screening the patient for cognitive and behavioral impairment (eg patient declines to be screened for cognitive or behavioral impairment) • Documentation of a system reason for not screening the patient for cognitive and behavioral impairment (eg no insurance to cover screening cost)
Supporting Guideline & Other References	**The following clinical recommendation statements are quoted verbatim from the referenced clinical guidelines and represent the evidence base for the measure:** • Screening for cognitive and behavioral impairment should be considered in patients with ALS (Level B).[1] • Screening tests of executive function may be considered to detect cognitive impairment in patients with ALS prior to confirmation with formal neuropsychological evaluation (Level C).[1] [1]Miller RG, Jackson CE, Kasarskis EJ, et al. Practice Parameter update: The care of the patient with amyotrophic lateral sclerosis: Multidisciplinary care, symptom management, and cognitive/behavioral impairment (an evidence-based review): Report of the Quality Standards Subcommittee of the American Academy of Neurology. *Neurology* 2009; 73:1227-1233

Measure Importance

Relationship to desired outcome

There is now considerable evidence for cognitive and behavioral manifestations in ALS. Specific ALS phenotypes include pure motor degeneration (ALS), ALS with cognitive impairment (ALSci), ALS with behavioral impairment (ALSbi), and ALS with a dementia meeting the Neary criteria for frontotemporal dementia (FTD) (ALS-FTD). FTD, as defined by Neary et al., has insidious onset, gradual progression, altered social conduct, emotional blunting, and loss of insight.[1] These criteria are required for the diagnosis of FTD, which is supported by neuropsychological abnormalities, language dysfunction, and poor self-care. ALSci reflects frontotemporal dysfunction with deficits in attention, cognitive flexibility, and word generation, with relative sparing of visuospatial function and memory. ALSbi refers to changes in social interactions unrelated to a psychiatric condition. The domain of cognitive

and behavioral impairment in ALS is a rapidly evolving field and there is emerging consensus regarding diagnostic criteria and assessment methods.[2] The ALS Cognitive Behavioral Screen (CBS) is specific to ALS, is validated, and has an accuracy of 100% to detect FTD, and 85% sensitivity to detect any cognitive impairment.[3]

Estimates of cognitive impairment range from 10%[2] to 75% [3] in those diagnosed with ALS. A population-based sample produced an estimate of 28%.[4] The prevalence of impairment meeting criteria for dementia ranged from 15%[5] to 41%.[6] Behavioral impairment (irritability and social disinhibition) was identified in 39%.[7] Three studies[7-9] documented mild cognitive decline over 6 months, while others[10,11] found no change over 12 months. It is not known whether patients can progress from ALSci or ALSbi to ALS-FTD. However, 15% of patients presenting with only FTD later develop motor neuron degeneration.[12]

A fuller characterization of the extent of cognitive and behavioral dysfunction in ALS has important implications given that the burden and stress for carers of patients with FTD is very great. It also has relevance to effective communication, legal issues and end-of-life decision making by patients with MND.[13] Recent studies have documented the utility of screening instruments in a busy ALS clinic.[14,15]Patients with cognitive and behavioral impairment were less compliant with management recommendations and had shorter survival.[16,17]

References

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Opportunity for Improvement	Cognitive impairment in ALS is best identified through neuropsychological assessment using standardized measures and normative data. The Mini-Mental State Examination is less sensitive to the cognitive impairment seen in ALS and does not examine for behavioral dysfunction. There is no consensus regarding the best screening tests for cognitive impairment in ALS. Two 1-minute word generation tests had 65% sensitivity, 90% specificity, and 88% positive predictive value in detecting possible, probable, or definite FTD by Neary criteria.[1] A 1-minute letter fluency measure (F words) had 73% sensitivity, 88% specificity, and 79% accuracy to detect ALSci.[2] An abbreviated neuropsychological battery demonstrated 88% accuracy.[2] Although there has not been a systematic study of how many clinics do screening, there is good support for the presence of a gap.[3] Recent studies have demonstrated the feasibility of screening patients in a busy specialized ALS clinic[4,5], but this is still not routinely practiced. A recent large study found that most patients and caregivers were not informed about the presence of cognitive or behavioral/ psychological impairment. Patients commonly reported being told by their doctor about physical symptoms such as problems walking (85%) or stiffness/cramps (74%) but not psychological issues like emotional liability (46%) or cognitive change (11%). Patients and caregivers have indicated that they do want to know about whether they are so affected. These data suggest that screening is not being done widely at all.[3] References [1]Rakowicz WP, Hodges JR. Dementia and aphasia in motor neuron disease: an under-recognized association? *J Neurol Neurosurg Psychiatry* 1998; 65:881– 889. [2] Lomen-Hoerth C, Murphy J, Langmore S Kramer JH, Olney RK, Miller B. Are amyotrophic lateral sclerosis patients cognitively normal? Neurology 60(7),1094-1097 (2003). [3] Wicks P, Frost J. ALS patients request more information about cognitive symptoms. *European J Neurol* 2008; 15:497-500. [4] Woolley SC, Utility of the ALS Cognitive Behavioral Screen. *Neurodegen Dis Manage* 2011;1:473-479. [5]Gordon PH, Wang Y, Doorish C, et al. A screening assessment of cognitive impairment in patients with ALS. *Amytroph Laterl Scler* 2007; 8:362-5
IOM Domains of Health Care Quality Addressed	Effective Patient-centered
Exclusion Justification	A medical reason exclusion has been included so that clinicians can exclude patients for whom a cognitive assessment may not be appropriate (eg, patient currently diagnosed with severe cognitive impairment). A patient reason exclusion has been included for patients who decline to screened for cognitive or behavioral impairment. A system reason exclusion has been included for patients who have no insurance to cover the screening cost.
Harmonization with Existing Measures	There are no other measures currently available that are similar to this measure or need to be harmonized with this measure.

Measure Designation

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

Level of Measurement	• Individual practitioner
Care setting	• Ambulatory Care
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

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 –CM Diagnosis Codes: 335.20 (amyotrophic lateral sclerosis) AND CPT E/M Service Code: 99201, 99202, 99203, 99204, 99205 (office-new patient), 99211,99212, 99213, 99214, 99215 (office-established patient), 99241, 99242, 99243, 99244, 99245 (outpatient consult), 99304, 99305, 99306, 99307, 99308, 99309, 99310 (nursing facility), 99324, 99325, 99326, 99327, 99328, 99334, 99335, 99336, 99337 (domiciliary), 99341, 99342, 99343, 99344, 99345, 99347, 99348, 99349, 99350 (home visit)
Numerator	Patients who are screened at least once annually for cognitive impairment (eg frontotemporal dementia screening or ALS Cognitive Behavioral Screen (CBS)) and behavioral impairment (eg ALS CBS). Reporting Instructions: • For all patients meeting the denominator criteria, report the CPT Category II, **3755F,** *Cognitive and behavioral impairment screening performed.* 3755 *Cognitive and behavioral impairment screening performed*
Denominator Exclusions	All patients with a diagnosis of amyotrophic lateral sclerosis. • Documentation of a medical reason for not screening the patient for cognitive and behavioral impairment (eg patient near end of life) Reporting Instructions: o For patient with appropriate exclusion, report **3755-1P** • Documentation of a patient reason for not screening the patient for cognitive and behavioral impairment (eg patient declines to be screened for cognitive or behavioral impairment) Reporting Instructions: o For patient with appropriate exclusion, report: **3755-2P**

- Documentation of a system reason for not screening the patient for cognitive and behavioral impairment (eg patient no insurance to cover screening cost)
  Reporting Instructions:
  - For patient with appropriate exclusion, report: **3755F-3P**

Measure #4: ALS Symptomatic Therapy Treatment Offered

Amyotrophic Lateral Sclerosis

Measure Description

Percentage of visits for patients with a diagnosis of ALS with patient offered treatment* for pseudobulbar affect, sialorrhea, and ALS related symptoms**.

*ALS treatment examples: eg dextromethorphan/quinidine combination, amitriptyline or fluoxetine for pseudobulbar affect; anti-inflammatory and analgesic agents for pain; anticholinergic agents for sialorrhea; botulinum toxin for refractory sialorrhea; tizanidine or baclofen for spasticity; antidepressants for depression; physical therapy for cramps; occupational therapy for adapted devices; or a dietary modification for constipation.
**ALS related symptoms definition: eg spasticity, muscle cramps, pain, anxiety, depression, leg swelling, insomnia, fatigue, laryngospasm or constipation

Measure Components

Numerator Statement	Patient visits with patient offered treatment* for pseudobulbar affect, sialorrhea, or ALS related symptoms**, if present. *ALS treatment examples: eg dextromethorphan/quinidine combination, amitriptyline or fluoxetine for pseudobulbar affect; anti-inflammatory and analgesic agents for pain; anticholinergic agents for sialorrhea; botulinum toxin for refractory sialorrhea; tizanidine or baclofen for spasticity; antidepressants for depression; physical therapy for cramps; occupational therapy for adapted devices; or a dietary modification for constipation. **ALS related symptoms definition: eg spasticity, muscle cramps, pain, anxiety, depression, leg swelling, insomnia, fatigue, laryngospasm or constipation
Denominator Statement	All visits for patients with a diagnosis of amyotrophic lateral sclerosis.
Denominator Exclusions	• No exclusions applicable for this measure.
Supporting Guideline & Other References	**The following clinical recommendation statements are quoted verbatim from the referenced clinical guidelines and represent the evidence base for the measure:** • In patients with ALS who have medically refractory sialorrhea, botulinum toxin B should be considered (Level B) and low-dose radiation therapy to the salivary glands may be considered (Level C).[1] • Provide a portable mechanical home suction device.[2] • Treat cramps in ALS with physiotherapy, physical exercise, and/or hydrotherapy.[2] • Antispasticity drugs such as baclofen and tizanidine may be tried.[2] • Physical therapy should be available regularly when there is significant spasticity.[2] [1]Miller RG, Jackson CE, Kasarskis EJ, et al. Practice Parameter update: The care of the patient with amyotrophic lateral sclerosis: Multidisciplinary care, symptom management, and cognitive/behavioral impairment (an evidence-based review): Report of the Quality Standards Subcommittee of the American Academy of Neurology. *Neurology* 2009; 73:1227-1233 [2]Andersen PM, Borasio GD, Dengler R, et al. EFNS task force on management of amyotrophic lateral sclerosis: guidelines for diagnosing and clinical care of patients and relatives. *European J of Neurology* 2005; 12:921-938 (GPP=Good Practice Point)

Measure Importance

Relationship to desired outcome

Pseudobulbar affect (PBA), excessive laughing or crying, or involuntary emotional expression disorder affects 20%–50% of patients with ALS, especially in pseudobulbar palsy.[1] Although it is not a mood disorder, antidepressants are frequently employed. Patients are embarrassed and isolated by these symptoms, which in turn greatly diminishes the patients' quality of life. A fixed-dose combination of dextromethorphan (DM)/quinidine (Q) (30 mg DM/30 mg Q BID) for treatment of pseudobulbar affect in ALS (Class I)[2] reduced the frequency and severity of laughing and crying behaviors compared to either DM (p<0.001) or Q alone (p<0.001). Side effects were dizziness, nausea, and somnolence, which accounted for termination of treatment in 24% with DM/Q compared to 6% with DM and 5% with Q. This formulation of DM/Q was not approved by the US Food and Drug Administration (FDA).

In a more recent study of 326 randomized patients, the PBA-episode daily rate was 49% (p<0.0001) lower for DM/Q-20/10 than for placebo[3]. Other endpoints showing statistically significant DM/Q benefit included the likelihood of PBA remission during the final 14 days. The low dosage was safe and well tolerated. On the basis of these two trials, the lower dose DM/Q-20/10 was recently approved by the US Food and Drug Administration (FDA) for the treatment of PBA.

Sialorrhea, or drooling, is embarrassing, socially isolating, and is associated with aspiration pneumonia. The prevalence is estimated at 50%, and 70% of patients receiving oral medications, mainly oral anticholinergic agents, for treatment reported benefit (Class III).[4,5] A portable mechanical home suction device is often helpful.[5] In a small trial, amitriptyline and botulinum toxin type A (BTxA) seemed equally effective, although 3 of 5 patients treated with amitriptyline experienced side effects (Class III).[6] In a double-blind, controlled trial of botulinum toxin type B (BTxB) in 20 patients with ALS with refractory sialorrhea (Class I)[7], patients were randomized to 2,500 U of BTxB or placebo into bilateral parotid and submandibular glands. Treated patients reported a global improvement of 82% at 2 and 4 weeks compared to 38% in placebo (p=0.05). At 12 weeks, 50% of patients receiving BTxB were improved compared to 14% receiving placebo. There were no important adverse events. Radiation therapy for medically refractory sialorrhea reduced salivary production, but side effects included erythema, sore throat, and nausea (Class III).[8] A "satisfactory response" was observed and saliva secretion rate diminished with a single dose of 7–7.5 Gy bilaterally (Class III).[9]

Fatigue may be a symptom of depression, poor sleep, abnormal muscle activation, immobility, or respiratory dysfunction. Fatigue diminishes quality of life for patients with ALS. Fatigue is a major issue for patients with ALS, and its etiology may be multifactorial (eg depression, anxiety, disturbed sleep, dyspnea). No single treatment is likely to be effective.[10,11] Fatigue was a side effect of therapy in 26% of patients taking riluzole vs. 13% taking placebo (p =0.07; number needed to harm=8) (Class III).[12] Asthenia occurred in 18% of patients taking riluzole vs. 12% of patients taking placebo in a larger study (p =0.004; number needed to harm=17) (Class III).[13]

Treating spasticity might improve gait and relieve painful spasms. Moderate exercise led to a small decline in the Ashworth Spasticity Scale over 3 months, compared to a worsening with no exercise (p < 0.005) (Class III).[14] Physical therapy is the mainstay of treatment of spasticity in ALS, and has been shown to be effective in a Class II study.[5]

The prevalence of depression in ALS ranges from 0 to 44%, although systematic studies suggest 10% in advanced ALS (Class III).[5,15] Depression shortens survival and lowers quality

of life for patients with ALS.[16] There is consensus among experts that depression should be treated in patients with ALS[5]; however, there are no controlled studies of benefit or harm.

Insomnia is common in ALS and may be a symptom of early respiratory weakness, underlying anxiety, depression, or pain.[17] There is a concern that sedative/hypnotic agents may suppress respiratory drive, tidal volume, and upper airway muscle tone in patients with ALS.

Laryngospasm is common in ALS and quite frightening to patients and families. Education about the benign nature of the symptom and its management is important.[11]

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Opportunity for Improvement

Quality of life is diminished by pseudobulbar affect, sialorrhea, and ALS related symptoms. Most of these symptoms are treatable, at least to some degree and could improve the patient's quality of life. Treatment for both refractory sialorrhea and pseudobulbar affect are underutilized in ALS.[1,2] Many practitioners are unaware of the condition and also of the recently approved treatment for pseudobulbar affect.[3]

Most other symptoms of ALS are treatable, albeit with less evidence. Still, most symptoms in ALS can be treated, as described above, and studies suggest that these treatments are underutilized.[1,2]

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IOM Domains of Health Care Quality Addressed	Effective Patient centered Timely
Exclusion Justification	No exclusions applicable for this measure.
Harmonization with Existing Measures	There are no other measures currently available that are similar to this measure or need to be harmonized with this measure.

Measure Designation

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

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 –CM Diagnosis Codes: 335.20 (amyotrophic lateral sclerosis)

	AND CPT E/M Service Code: 99201, 99202, 99203, 99204, 99205 (office-new patient), 99211,99212, 99213, 99214, 99215 (office-established patient), 99241, 99242, 99243, 99244, 99245 (outpatient consult), 99304, 99305, 99306, 99307, 99308, 99309, 99310 (nursing facility), 99324, 99325, 99326, 99327, 99328, 99334, 99335, 99336, 99337 (domiciliary), 99341, 99342, 99343, 99344, 99345, 99347, 99348, 99349, 99350 (home visit)
Numerator	Patient visits with patient offered treatment* for pseudobulbar affect, sialorrhea, or ALS related symptoms**, if present. *ALS treatment examples: eg dextromethorphan/quinidine combination, amitriptyline or fluoxetine for pseudobulbar affect; anti-inflammatory and analgesic agents for pain; anticholinergic agents for sialorrhea; botulinum toxin for refractory sialorrhea; tizanidine or baclofen for spasticity; antidepressants for depression; physical therapy for cramps; occupational therapy for adapted devices; or a dietary modification for constipation. **ALS related symptoms definition: eg spasticity, muscle cramps, pain, anxiety, depression, leg swelling, insomnia, fatigue, laryngospasm or constipation Reporting Instructions: • For all patients meeting denominator criteria, report either **3756F**, *Patient has pseudobulbar affect, sialorrhea or ALS related symptoms* **OR 3757F,** *Patient does not have pseudobulbar affect, sialorrhea, or ALS related symptoms*. • When **3756F** is reported, also report **4541F,** *Patient offered treatment for pseudobulbar affect, sialorrhea, or ALS related symptoms.* 3756F-*Patient has pseudobulbar affect, sialorrhea, or ALS related symptoms* 3757F-*Patient does not have pseudobulbar affect, sialorrhea, or ALS related symptoms* 4541F-*Patient offered treatment for pseudobulbar affect, sialorrhea, or ALS related symptoms*
Denominator Exclusions	All visits for patients with a diagnosis of amyotrophic lateral sclerosis. Reporting Instructions: • There are no exclusions for this measure. Do not report modifiers 1P, 2P or 3P with 4541F.

Measure #5: ALS Respiratory Insufficiency Querying and Referral for Pulmonary Function Testing

Amyotrophic Lateral Sclerosis

Measure Description

Percentage of patients with a diagnosis of amyotrophic lateral sclerosis who were queried about symptoms of respiratory insufficiency (awake or associated with sleep) and referred for pulmonary function testing (eg vital capacity (VC), maximum inspiratory pressure (MIP), sniff nasal pressure (SNP), or peak cough expiratory flow (PCEF)), at least every three months.

Measure Components

Numerator Statement	Patients who were who were queried about symptoms of respiratory insufficiency (awake or associated with sleep) and referred for pulmonary function testing (eg vital capacity (VC), maximum inspiratory pressure (MIP), sniff nasal pressure (SNP), or peak cough expiratory flow (PCEF)), at least every three months.
Denominator Statement	All patients with a diagnosis of amyotrophic lateral sclerosis.
Denominator Exclusions	• Documentation of medical reason for not querying about symptoms of respiratory and referring for pulmonary function testing or peak cough expiratory flow (eg patient with severe cognitive impairment who cannot answer any queries) • Documentation of patient reason for not querying about symptoms of respiratory and referring for pulmonary function testing or peak cough expiratory flow (eg patient declines to be referred for pulmonary function testing)
Supporting Guideline & Other References	**The following clinical recommendation statements are quoted verbatim from the referenced clinical guidelines and represent the evidence base for the measure:** • Symptoms or signs of respiratory insufficiency should initiate discussions with the patient and the caregivers about all treatment options such as noninvasive ventilation (NIV), tracheostomy ventilation (TV) and the terminal phase. Early discussions are needed to allow advance planning and directives. The patient should be informed about the temporary nature of NIV (which is primarily directed towards improving quality of life rather than prolonging it (as opposed to TV)). Care should adapt to the changing needs of patients and carers over the course of the disease. (GPP)[1] • Vital capacity (VC) is the most available and practical test for the monitoring of respiratory function on a regular basis. If possible, VC should be measured both standing/sitting and lying. (GPP)[1] • Supine FVC and MIP may be considered useful in routine respiratory monitoring, in addition to the erect FVC (Level C).[2] • Sniff nasal pressure (SNP) may be considered to detect hypercapnia and nocturnal hypoxemia (Level C).[2] • SNP may be used for monitoring of inspiratory muscle strength, particularly in some bulbar patients who cannot perform VC accurately. (GPP)[1] • Nocturnal oximetry, available at home, is recommended in patients with symptoms of nocturnal hypoventilation. (GPP)[1] • Nocturnal oximetry may be considered to detect hypoventilation (regardless of the forced vital capacity (FVC)). (Level C)[2]

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Measure Importance

Relationship to desired outcome

Patients should be queried about symptoms of respiratory insufficiency, screened with pulmonary function testing on a regular basis, and referred for pulmonary consultation when appropriate. Treatment of respiratory insufficiency improves survival, quality of life and respiratory symptoms.[1,2] The diagnosis and management of respiratory insufficiency is critical because most deaths from ALS are due to respiratory failure. Published guidelines for respiratory care were based on clinical experience, expert opinion, and observational research.[1-4]

FVC is the most commonly used respiratory measurement in ALS[5] and it was a significant predictor of survival.[6] FVC may be an insensitive measure of respiratory muscle strength since 13/20 patients with an FVC >70% had abnormal maximal inspiratory pressure (MIP) <−60 cm.[7]

Nocturnal desaturations <90% for 1 cumulative minute was a more sensitive indicator of nocturnal hypoventilation than either FVC or MIP.[7] FVC correlated poorly with symptoms of nocturnal hypoventilation and desaturation.[8] Nocturnal oximetry correlated with survival.[9]

Supine FVC, although more difficult to perform, may be a better predictor of diaphragm weakness than erect FVC. FVC closely correlated with transdiaphragmatic pressure (Pdi), and a supine FVC <75% reliably predicted an abnormally low Pdi.[10] Further, the difference between erect and supine FVC correlated with orthopnea.[11]

The sniff transdiaphragmatic pressure (sniff Pdi) detected hypercapnia with a sensitivity of 90% and a specificity of 87%. Sniff nasal pressure (SNP) showed greater predictive power of survival than either FVC or MIP. When SNP was less than 30 cm, median survival was 3 months. In addition, SNP was more reliably recorded at later stages of ALS than either FVC or MIP.[1,3-4]

The peak cough expiratory flow (PCEF) remains the most widely used measure of cough effectiveness. Patients with a mean PCEF above 337 L/min had a significantly greater chance of being alive at 18 months.[1]

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2009;73:1218-1226
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Opportunity for Improvement

The vast majority of patients with ALS only get standard vital capacity measures to monitor pulmonary function, and in many instances even this basic measurement is not made.[1,2,6] The use of nocturnal oximetry, polysomnography, sniff nasal pressure (SNP), maximum inspiratory pressure (MIP), peak expiratory cough flow (PCEF) and supine vital capacity (VC) should permit earlier detection and treatment of respiratory insufficiency in ALS.[1-3] More studies are needed to clarify which of these measures is the best, but several studies have shown that MIP is probably the most sensitive indicator of early diaphragm weakness.[3-5]

Symptoms of early respiratory insufficiency should be specifically asked for: dyspnea, orthopnea, excessive daytime sleepiness, insomnia, fatigue, morning headache. Patients often do not volunteer these symptoms and the clinician may detect early respiratory insufficiency by enquiring about them, which is often not done.[2,3,6]

References

[1]Miller RG, Jackson CE, Kasarskis EJ, et al. Practice Parameter Update: The care of the patient with amyotrophic lateral sclerosis: Drug, nutritional, and respiratory therapies (an evidence-based review): Report of the Quality Standards Subcommittee of the American Academy of Neurology *Neurology* 2009;73:1218-1226
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IOM Domains of Health Care Quality Addressed	Effective Patient centered
Exclusion Justification	A medical reason exclusion for patients with severe cognitive impairment who cannot answer any queries. A patient reason exclusion has been included for patients who decline to be referred for pulmonary function testing.
Harmonization with Existing Measures	There are no other measures currently available that are similar to this measure or need to be harmonized with this measure.

Measure Designation

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

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 –CM Diagnosis Codes: 335.20 (amyotrophic lateral sclerosis) AND CPT E/M Service Code: 99201, 99202, 99203, 99204, 99205 (office-new patient), 99211,99212, 99213, 99214, 99215 (office-established patient), 99241, 99242, 99243, 99244, 99245 (outpatient consult), 99304, 99305, 99306, 99307, 99308, 99309, 99310 (nursing facility), 99324, 99325, 99326, 99327, 99328, 99334, 99335, 99336, 99337 (domiciliary), 99341, 99342, 99343, 99344, 99345, 99347, 99348, 99349, 99350 (home visit).
Numerator	Patients who were who were queried about symptoms of respiratory insufficiency (awake or associated with sleep) and referred for pulmonary function testing (eg vital capacity (VC), maximum inspiratory pressure (MIP), sniff nasal pressure (SNP), or

peak cough expiratory flow (PCEF)), at least every three months.

Reporting Instructions:

- For all patients meeting denominator criteria, report the CPT Category II **1503F,** *Patient queried about symptoms of respiratory insufficiency* **AND 3758F**, *Patient referred for pulmonary function testing or peak cough expiratory flow*.

1503F-*Patient queried about symptoms of respiratory insufficiency*
3758F-*Patient referred for pulmonary function testing or peak cough expiratory flow*

Denominator Exclusions

All patients with a diagnosis of amyotrophic lateral sclerosis.

- Documentation of medical reason for not querying about symptoms of respiratory insufficiency and referring for pulmonary function testing or peak cough exploratory flow (eg patient with severe cognitive impairment who cannot answer the query)
  Reporting Instructions:
  - For patient with appropriate exclusion criteria, report: **1503F-1P AND 3758F-1P**

- Documentation of patient reason for not querying about symptoms of respiratory insufficiency and referring for pulmonary function testing or peak cough expiratory flow (eg patient declines to be referred)
  Reporting Instructions:
  - For patient with appropriate exclusion criteria, report: **1503F-2P AND 3758F-2P**

Measure #6: ALS Noninvasive Ventilation Treatment for Respiratory Insufficiency Discussed

Amyotrophic Lateral Sclerosis

Measure Description

Percentage of patients diagnosed with ALS and respiratory insufficiency with whom the clinician discussed at least once annually treatment options for noninvasive respiratory support (eg noninvasive ventilation (NIV), assisted cough).

Measure Components

Numerator Statement	Patients with whom the clinician discussed at least once annually treatment options for noninvasive respiratory support (eg noninvasive ventilation (NIV), assisted cough).
Denominator Statement	All patients with a diagnosis of amyotrophic lateral sclerosis and respiratory insufficiency.
Denominator Exclusions	• Documentation of a medical reason for not discussing treatment options for noninvasive respiratory support (eg patient is in a coma; patient has severe cognitive impairment and cannot communicate; patient is already on appropriate respiratory support) • Documentation of patient reason for not discussing treatment options for noninvasive respiratory support (eg patient declines to discuss treatment options)
Supporting Guideline & Other References	**The following clinical recommendation statements are quoted verbatim from the referenced clinical guidelines and represent the evidence base for the measure:** • NIV should be considered to treatment respiratory insufficiency in ALS, both to lengthen survival and to slow the rate of FVC decline (Level B).[1] • Symptoms or signs of respiratory insufficiency should initiate discussions with the patient and the caregivers about all treatment options such as NIV, TV and support in the terminal phase. Early discussions are needed to allow advance planning and directives.[2] • NIV may be considered to enhance QOL in patients with ALS who have respiratory insufficiency (Level C).[1] • NIV may be considered at the earliest sign of nocturnal hypoventilation or respiratory insufficiency in order to improve compliance with NIV in patients with ALS (Level C).[1] • Use non-invasive ventilation or invasive ventilation for alleviating breathlessness (Level II). (Caution is recommended in the use of oxygen therapy to supplement ventilator support as there is no evidence that it is advantageous to MND patients.)[3] • Use non-invasive intermittent positive pressure ventilators (NIPPV) or bi-level positive airway pressure ventilators to provide non-invasive ventilation, depending upon local availability and expertise, since both have been shown to improve quality of life and survival in MND/ALS patients (Level II).[3] • Consider non-invasive ventilation as an initial intervention in patients with, or at risk of developing, hypercapnia. (Grade D)[4] • Offer a trial of non-invasive ventilation if the patient's symptoms and signs and the results of the respiratory function tests indicate that the patient is likely to benefit from the treatment.(Low Quality*)[5] • When oxygen saturation falls below 95% the use of noninvasive ventilation and/or strategies to aid airway clearance should be considered. (Grade D)[4]

	• Oxygen therapy should be avoided or used with extreme caution due to the risk of carbon dioxide retention in patients with neuromuscular disease. (Grade A)[4] • The patient should be informed about the temporary nature of NIV. Care should adapt to the changing needs of patients and carers over the course of the disease. (GPP)[2] • TIV may be considered to preserve QOL in patients with ALS who want long-term ventilator support (Level C).[1] • Mechanical Insufflation/exsufflation (MIE) may be considered to clear secretions in patients with ALS who have reduced peak cough flow, particularly during an acute chest infection (Level C).[1] [1]Miller RG, Jackson CE, Kasarskis EJ, et al. Practice Parameter Update: The care of the patient with amyotrophic lateral sclerosis: Drug, nutritional, and respiratory therapies (an evidence-based review): Report of the Quality Standards Subcommittee of the American Academy of Neurology *Neurology* 2009; 73:1218-1226 [2]Andersen PM, Borasio GD, Dengler R, et al. EFNS task force on management of amyotrophic lateral sclerosis: guidelines for diagnosing and clinical care of patients and relatives. *European J of Neurology* 2005;12:921-938 (GPP=Good Practice Point) [3]Heffernan CC, Jenkinson, T. Holmes, et al. Management of respiration in MND/ALS patients: An evidence based review. *Amyotrophic Lateral Sclerosis* 2006; 7(1):5-15. [4]Bott J, Blumenthal S, Buxton M, et al. Guidelines for the physiotherapy management of the adult, medical, spontaneously breathing patient. *Thorax* 2009; 64:1-51. [5] National Institute for Health and Clinical Excellence (2010) Motor neurone disease: the use of non-invasive ventilation in the management of motor neurone disease. London: National Institute for Health and Clinical Excellence. Available from: www.nice.org.uk/guidance/CG105 * NICE downgraded the level of evidence to low quality without citing an appropriate reason for doing so.

Measure Importance

Relationship to desired outcome	There is evidence that implementation of noninvasive ventilation (NIV) will result in improved survival. In a randomized controlled study, patients using NIV experienced a median survival benefit of 205 days (Class I).[1] NIV was initiated based on orthopnea with an MIP $\leq$60 cm or symptomatic hypercapnia. No survival benefit was seen in patients with poor bulbar function. "Early" intervention with NIV (nocturnal oximetry demonstrating >15 desaturation events/hour) resulted in 11 months longer survival compared to controls, with some beneficial effect in bulbar patients (Class III).[2] Patients who used NIV >4 hours/day survived 7 months longer than patients using the device <4 hours/day (Class III).[3] FVC declined more slowly after introducing NIV (pre -2.2%/month compared to post -1.1%/ month) (Class I/III)[2] and the decline was slower in those who used NIV > 4 hours/day (Class III).[3] A survival benefit of 20 months was observed in NIV tolerant patients vs. 5 months in NIV-intolerant patients (Class III).[4] NIV is probably effective in prolonging survival (1 Class I, 3 Class III studies) and in slowing the rate of FVC decline (1 Class I, 1 Class III study). References [1]Bourke SC, Tomlinson M, Williams TL, et al. Effects of non-invasive ventilation on survival and quality of life in patients with amyotrophic lateral sclerosis: a randomised controlled trial. *Lancet Neurol* 2006; 5:140 –147. [2] Pinto A, de Carvalho M, Evangelista T, et al. Nocturnal pulse oximetry: a new approach to establish the appropriate time for non-invasive ventilation in ALS patients. *Amyotroph Lateral Scler Other Motor*

Neuron Disord 2003; 4(1):31-5.
[3] Kleopa KA, Sherman M, Neal B, et al. Bipap improves survival and rate of pulmonary function decline in patients with ALS. *J Neurol Sci.* 1999; 164(1):82-8.
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Opportunity for Improvement	Survival is improved with increased proportion of use of noninvasive ventilation from 16% of ALS patients with respiratory insufficiency to 51%[1-2], while over 65% of ALS patients have said that they would want respiratory support indicating that noninvasive ventilation is still underutilized in ALS patients.[3-5] Even though published guidelines to date have made a positive impact in doubling utilization rates, underutilization of this important treatment is still evident.[3,4] Moreover, recent studies document frequent nocturnal patient-ventilator asynchrony in patients with ALS, even when using NIV prescribed as per current AAN practice parameters.[6] These findings suggest that use of NIV per these current parameters is unlikely to provide patients with ALS optimal nocturnal ventilatory support. More attention to monitoring the quality of patient sleep and oxygenation with NIV, and reducing asynchrony, is likely to improve the efficacy of this treatment.[6]

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IOM Domains of Health Care Quality Addressed	Effective Patient centered
Exclusion Justification	A medical reason exclusion has been included for patients who cannot communicate (eg patient in a coma or who has severe cognitive impairment) or who are already on appropriate respiratory support. A patient reason exclusion has been for patients who to decline to discuss treatment options.
Harmonization with Existing Measures	There are no other measures currently available that are similar to this measure or need to be harmonized with this measure.

Measure Designation

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

Level of Measurement	• Individual practitioner
Care setting	• Ambulatory Care
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

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 –CM Diagnosis Codes: 335.20 amyotrophic lateral sclerosis 518.83 Respiratory Insufficiency **AND** CPT E/M Service Code: 99201, 99202, 99203, 99204, 99205 (office-new patient), 99211,99212, 99213, 99214, 99215 (office-established patient), 99241, 99242, 99243, 99244, 99245 (outpatient consult), 99304, 99305, 99306, 99307, 99308, 99309, 99310 (nursing facility), 99324, 99325, 99326, 99327, 99328, 99334, 99335, 99336, 99337 (domiciliary), 99341, 99342, 99343, 99344, 99345, 99347, 99348, 99349, 99350 (home visit).
Numerator	Patients with whom the clinician discussed at least once annually treatment options for noninvasive respiratory support (eg noninvasive ventilation (NIV), assisted cough). Reporting Instructions: • For all patients meeting denominator criteria, report the CPT Category II, **1504F** *Patient has respiratory insufficiency,* or **1505F**, *Patient does not have respiratory insufficiency*. • When **1504** is reported, also report **4XXXF3**, *Options for noninvasive respiratory support discussed with patient.* 1504F-*Patient has respiratory insufficiency* 1505F-*Patient does not have respiratory insufficiency* 4550F-*Options for noninvasive respiratory support discussed with patient*
Denominator Exclusions	All patients with a diagnosis of amyotrophic lateral sclerosis and respiratory insufficiency. • Documentation of a medical reason for not discussing treatment options for noninvasive respiratory support (eg patient is in a coma; patient has severe cognitive impairment and cannot communicate; patient is already on

appropriate respiratory support)
Reporting Instructions:
- For patient with appropriate exclusion criteria, report: **4550F-1P**

- Documentation of patient reason for not discussing treatment options for noninvasive respiratory support (eg patient declines to discuss treatment options)
  Reporting Instructions:
  - For patient with appropriate exclusion criteria, report:**4550F-2P**

Measure #7: ALS Screening for Dysphagia, Weight Loss or Impaired Nutrition

Amyotrophic Lateral Sclerosis

Measure Description

Percentage of patients diagnosed with ALS who were screened at least every 3 months for dysphagia, weight loss or impaired nutrition* and the result(s) of the screening(s) was documented in the medical record.

*Impaired nutrition includes: changes in nutritional biomarkers (serum prealbumin, total protein, or hemoglobin) or body mass index

Measure Components

Numerator Statement	Patients who were screened at least every 3 months for dysphagia, weight loss or impaired nutrition* and the result(s) of the screening(s) was documented in the medical record. *Impaired nutrition includes: changes in nutritional biomarkers (serum prealbumin, total protein, or hemoglobin) or body mass index
Denominator Statement	All patients with a diagnosis of amyotrophic lateral sclerosis.
Denominator Exclusions	• Documentation of a patient reason for not screening for dysphagia, weight loss or impaired nutrition and documenting the result(s) of the screening(s) in the medical record (eg patient declines screening) • Documentation of a system reason for not screening for dysphagia, weight loss or impaired nutrition and documenting the result(s) of the screening(s) in the medical record (eg equipment not available to complete the screenings; no insurance)
Supporting Guideline & Other References	**The following clinical recommendation statements are quoted verbatim from the referenced clinical guidelines and represent the evidence base for the measure:** • Bulbar dysfunction and nutritional status, including weight, should be checked at each visit.(GPP)[1] • The patient and spouse should be referred to a dietician as soon as dysphagia appears. A speech and language therapist (SLT) can give valuable advice on swallowing techniques.(GPP)[1] [1]Andersen PM, Borasio GD, Dengler R, et al. EFNS task force on management of amyotrophic lateral sclerosis: guidelines for diagnosing and clinical care of patients and relatives. *European J of Neurology* 2005; 12:921-938 (GPP=Good Practice Point)

Measure Importance

Relationship to desired outcome

Weight loss is a key prognostic indicator for ALS with the risk of death increased 7-fold when body mass index is <18.5 kg/m2. Criteria for gastrostomy are loss of >10% of usual bodyweight since the onset of disease (> 5% loss at diagnosis and > 5% loss from usual body weight at follow-up). Weight loss is related to bulbar involvement, upper limb disability, depression and/or hypermetabolism. Nutritional status should be checked at 3 month intervals by measuring weight, assessing frequency/severity of choking, duration of meals and caloric intake.[1-6] A speech language pathologist (SLP) helps manage dysphagia to lower the risk of aspiration and optimize oral intake.[7] Nutritional interventions before deployment of gastrostomy tube may include modification of the texture and consistency of food, and increased caloric intake. Hyperlipemia may significantly prolong survival in ALS but the value

of increasing lipid intake is unknown.[1-6]

In 9 studies, a total of 469 patients with ALS received enteral nutrition via PEG [8-16] Using patients as their own controls, 7 Class III studies demonstrated either weight stabilization or modest weight gain over 2–24 months.[8-10, 12, 13, 15,16] In 2 Class II studies[11, 14] in which PEG refusers served as controls, weight stabilization was demonstrated in the PEG group vs. continued weight loss in controls ($p \leq 0.03$). Enteral nutrition administered via PEG is probably effective in stabilizing body weight/body mass index (2 Class II, 7 Class III studies).[11,14]

References
[1] Marin B, Desport JC, Kajeu P, et al. Alteration of nutritional status at diagnosis is a prognostic factor for survival of amyotrophic lateral sclerosis patients. *J Neurol Neurosurg Psychiatry* 2011;82(6):628-34.
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[7] Kasarskis EJ, Scarlata D, Hill R, et al. A retrospective study of percutaneous endoscopic gastrostomy in ALS patients during the BDNF and CNTF trials. *J Neurol Sci* 1999;169:118 –125.
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[15] Mitsumoto H, Davidson M, Moore D, et al. Percutaneous endoscopic gastrostomy (PEG) in patients with ALS and bulbar dysfunction. *Amyotroph Lateral Scler Other Motor Neuron Disord* 2003; 4:177–185.
[16] Rozier A, Ruskone Fourmestraux A, Rosenbaum A, et al. Role of percutaneous endoscopic gastrostomy in amyotrophic lateral sclerosis [in French]. *Rev Neurol* 1991; 147:174 –176.

Opportunity for Improvement

The prevalence of malnutrition varies between 16 - 55% in ALS patients across several studies. [1-3] There is an adjusted 30% increased risk of death for a 5% decrease from usual weight at time of ALS diagnosis (RR 1.30; 95% CI 1.08 to 1.56). During follow-up, there is an adjusted 34% (95% CI 18% to 51%) and 24% (95% CI 13% to 36%) increased risks of death associated with each 5% decrease in usual weight and each unit decrease in usual BMI, respectively ($p<0.0001$). Malnutrition during the course of ALS was related to a shorter survival ($p=0.01$), and fat mass level was associated with a better outcome (RR 0.90 for each 2.5 kg fat mass increment). [2] Thus, many patients lose weight in ALS and survive for a shorter time.

Treatment to stabilize weight and lengthen survival, with nutritional supplements and enteral feeding, is underutilized. Only 19% of patients utilized nutritional supplements and only 16% of patients utilized enteral feeding in one large study.[4]

References
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[4]Miller RG, Anderson F, Brooks BR, set al. ALS CARE Study Group. Outcomes research in amyotrophic lateral sclerosis: lessons learned from the amyotrophic lateral sclerosis clinical assessment, research, and education database. *Ann Neurol*. 2009; 65 Suppl 1:S24-8.

IOM Domains of Health Care Quality Addressed	Effective Patient centered
Exclusion Justification	A patient reason exclusion has been included for patients who chose not to accept malnutrition screening. A system reason exclusion has been included for patients who have no insurance or resources to pay for malnutrition screening.
Harmonization with Existing Measures	There are no other measures currently available that are similar to this measure or need to be harmonized with this measure.

Measure Designation

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

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	ICD-9 –CM Diagnosis Codes:

(Eligible Population)	335.20 amyotrophic lateral sclerosis AND CPT E/M Service Code: 99201, 99202, 99203, 99204, 99205 (office-new patient), 99211,99212, 99213, 99214, 99215 (office-established patient), 99241, 99242, 99243, 99244, 99245 (outpatient consult), 99304, 99305, 99306, 99307, 99308, 99309, 99310 (nursing facility), 99324, 99325, 99326, 99327, 99328, 99334, 99335, 99336, 99337 (domiciliary), 99341, 99342, 99343, 99344, 99345, 99347, 99348, 99349, 99350 (home visit).
Numerator	Patients who were screened at least every 3 months for dysphagia, weight loss or impaired nutrition* and the result(s) of the screening(s) was documented in the medical record. *Impaired nutrition definition: changes in nutritional biomarkers (serum albumin, total protein, cholinesterase, or hemoglobin) or anthropometric measures (skinfold, muscle area) Reporting Instructions: • For all patients meeting denominator criteria, report the CPT Category II, **3759F,** *Patient screened for dysphagia, weight loss, and impaired nutrition, and results documented.* 3759F- *Patient screened for dysphagia, weight loss, and impaired nutrition, and results documented*
Denominator Exclusions	All patients with a diagnosis of amyotrophic lateral sclerosis. • Documentation of a patient reason for not screening for dysphagia, weight loss or impaired nutrition and documenting the result(s) of the screening(s) in the medical record (eg patient declines screening) Reporting Instructions: o For patient with appropriate exclusion criteria, report: **3759F-2P** • Documentation of a system reason for not screening for dysphagia, weight loss or impaired nutrition and documenting the result(s) of the screening(s) in the medical record (eg equipment not available to complete the screenings; no insurance). Reporting Instructions: o For patient with appropriate exclusion criteria, report: **3759F-3P**

Measure #8: ALS Nutritional Support Offered

Amyotrophic Lateral Sclerosis

Measure Description

Percentage of patients diagnosed with ALS and dysphagia, weight loss, or impaired nutrition who were offered at least once annually dietary or enteral nutrition support via PEG or RIG*.
*PEG-percutaneous endoscopic gastrostomy. RIG-radiographic inserted gastrostomy.

Measure Components

Numerator Statement	Patients who were offered at least once annually dietary or enteral nutrition support via PEG or RIG*. *PEG-percutaneous endoscopic gastrostomy. RIG-radiographic inserted gastrostomy
Denominator Statement	All patients with a diagnosis of amyotrophic lateral sclerosis and dysphagia, weight loss or impaired nutrition.
Denominator Exclusions	• Documentation of a medical reason for not offering dietary or enteral nutritional support (eg patient already on PEG/RIG; patient cannot tolerate the procedure)
Supporting Guideline & Other References	**The following clinical recommendation statements are quoted verbatim from the referenced clinical guidelines and represent the evidence base for the measure:** • In patients with ALS with impaired oral food intake, enteral nutrition via PEG/RIG should be considered to stabilize body weight (Level B).[1] • PEG or RIG should be considered for prolonging survival in patient with ALS. (Level B).[1] • When PEG is indicated, patient and carers should be informed: (i) of the benefits and risks of the procedure; (ii) that it is possible to continue to take food orally as long as it is possible; (iii) that deferring PEG to a late disease stage may increase the risk of the procedure. (GPP)[2] • The timing of PEG/RIG is based on an individual approach taking into account bulbar symptoms, malnutrition (weight loss >10%), respiratory function and the patient's general condition. Thus, early operation is highly recommended.(GPP)[2] [1]Miller RG, Jackson CE, Kasarskis EJ, et al. Practice Parameter Update: The care of the patient with amyotrophic lateral sclerosis: Drug, nutritional, and respiratory therapies (an evidence-based review): Report of the Quality Standards Subcommittee of the American Academy of Neurology *Neurology* 2009; 73:1218-1226 [2]Andersen PM, Borasio GD, Dengler R, et al. EFNS task force on management of amyotrophic lateral sclerosis: guidelines for diagnosing and clinical care of patients and relatives. *European J of Neurology* 2005; 12:921-938 (GPP=Good Practice Point)

Measure Importance

Relationship to desired outcome	There is supportive evidence that implementation of enteral feeding will result in improved survival. Two Class II and 7 Class III studies compared survival in patients receiving PEG (n=585) vs. those without PEG (n=1619). One Class III study demonstrated a survival advantage vs. control with multivariate analysis (p=0.02), but not with univariate analysis (p=0.09).[1] A Class III population-based study from Italy found improved survival with PEG compared to patients with oral intake, also based on a multivariate analysis (3.89-fold; p =

0.0004).[2] Two Class II studies demonstrated prolonged survival in the PEG group vs. PEG refusers similar results when patients with PEG were compared to nasogastric-fed controls (p=0.03) (Class III). However, 4 Class III studies failed to find a significant survival benefit with PEG.[1-4] All but one[6] of the negative studies included patients not needing PEG as a control group. The positive studies used controls that refused PEG (Class II) [7-8] or used a risk model and multivariate analysis based on factors that predicted survival (statistically controlling for confounders) (Class III).[1-2] Studies using appropriate controls or multivariate analysis demonstrated that PEG is probably effective in prolonging survival in ALS, although insufficient data exist to quantitate the survival advantage (2 Class II studies).[9]

References
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Opportunity for Improvement

Not all patients who should have enteral feeding receive this treatment with only 43% of ALS patients who met guideline indications for gastrostomy tube placement undergoing the procedure.[1] Increasing proportions of patients who would most likely benefit are now receiving treatment but not all.[2-5]

Poor respiratory function increases the risk of the procedure.[1] Early detection of impaired nutrition and referral to a gastroenterologist will increase the safety and utilization of PEG and RIG.[2-4]

References
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IOM Domains of Health Care Quality Addressed	Effective Patient centered Timely
Exclusion Justification	A medical reason exclusion has been included for patients who are already on PEG/RIG, or could not tolerate the procedure.
Harmonization with Existing Measures	There are no other measures currently available that are similar to this measure or need to be harmonized with this measure.

Measure Designation

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

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 –CM Diagnosis Codes: 335.20 amyotrophic lateral sclerosis 783.21 Loss of weight, 787.2 Dysphagia, 783.21 Impaired nutrition, 783.3 Feeding difficulties and mismanagement, or 263.9 for impaired nutrition AND CPT E/M Service Code: 99201, 99202, 99203, 99204, 99205 (office-new patient), 99211,99212, 99213, 99214, 99215 (office-established patient), 99241, 99242, 99243, 99244, 99245 (outpatient consult), 99304, 99305, 99306, 99307, 99308, 99309, 99310 (nursing facility), 99324, 99325, 99326, 99327, 99328, 99334, 99335, 99336, 99337 (domiciliary), 99341, 99342, 99343, 99344, 99345, 99347, 99348, 99349, 99350 (home visit),
Numerator	Patients who were offered at least once annually dietary or enteral nutrition support via PEG or RIG*.

	*PEG-percutaneous endoscopic gastrostomy. RIG-radiographic inserted gastrostomy Reporting Instructions: • For all patients meeting denominator criteria, report CPT Category II Code **3760F**, *Patient exhibits dysphagia, weight loss, or impaired nutrition* or **3761F**, *Patient does not exhibit dysphagia, weight loss, or impaired nutrition*. • When **3760F** is reported, also report **4551F,** *Nutritional support offered.* 3760F *Patient exhibits dysphagia, weight loss, or impaired nutrition* 3761F *Patient does not exhibit dysphagia, weight loss, or impaired nutrition* 4551F *Nutritional support offered*
Denominator Exclusions	All patients with a diagnosis of amyotrophic lateral sclerosis and dysphagia, weight loss or impaired nutrition. • Documentation of a medical reason for not offering dietary or enteral nutritional support at least once annually (eg patient already on PEG/RIG, patient cannot tolerate the procedure) Reporting Instructions: o For patient with appropriate exclusion criteria, report: **4551F-1P**

Measure #9: ALS Communication Support Referral

Amyotrophic Lateral Sclerosis

Measure Description

Percentage of patients diagnosed with amyotrophic lateral sclerosis who are dysarthric who were offered a referral at least once annually to a speech language pathologist for an augmentative/alternative communication evaluation.

Measure Components

Numerator Statement	Patients who were offered a referral at least once annually to a speech language pathologist for an augmentative/alternative communication evaluation.
Denominator Statement	All patients with a diagnosis of amyotrophic lateral sclerosis who are dysarthric.
Denominator Exclusions	• Documentation of a medical reason for not offering a referral to a speech language pathologist for an augmentative/alternative communication evaluation (eg patient is already using an augmentative communication device).
Supporting Guideline & Other References	**The following clinical recommendation statements are quoted verbatim from the referenced clinical guidelines and represent the evidence base for the measure:** • Regular assessment (i.e. every 3-months) of communication by a trained speech therapist is recommended. (GPP)[1] • The use of appropriate communication support systems (ranging from pointing boards with figures or words, to computerized speech synthesizers) should be provided as required. (GPP)[1] [1]Andersen PM, Borasio GD, Dengler R, et al. EFNS task force on management of amyotrophic lateral sclerosis: guidelines for diagnosing and clinical care of patients and relatives. *European J of Neurology* 2005;12:921-938 (GPP=Good Practice Point)

Measure Importance

Relationship to desired outcome

Communication is vital to quality of life and most ALS patients lose the ability to communicate.[1,2] Both high tech and low tech options are available through a speech language pathologist to enhance continued communication.[3] Dysarthria is present in nearly all ALS patients with bulbar onset and in nearly 70% of ALS patients with spinal onset. More than 95% of ALS patients cannot speak before death and patients who accept gastrostomy tube, non-invasive ventilation or tracheostomy-assisted ventilation have a greater need for augmentative alternative communication as the disease progresses.[1-3]

References

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Opportunity for Improvement	Speech assessment in ALS patients identifies dysarthria, independent of dysphagia, that limits communication and maintenance of their active communicator role. Speech correction should focus on the maintenance of functional communication. Assessment should involve measurement of speech rate (words per minute). Augmentative and alternative communication is recommended when speech rate is less than 125 words per minute. Nearly 88% of ALS patients are evaluated by this criterion, but fewer than half implement appropriate interventions. [1-5]

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IOM Domains of Health Care Quality Addressed	Effective Patient centered
Exclusion Justification	A medical reason exclusion has been included for patients who are already using an augmentative communication device.
Harmonization with Existing Measures	There are no other measures currently available that are similar to this measure or need to be harmonized with this measure.

Measure Designation

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

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 –CM Diagnosis Codes: 335.20 amyotrophic lateral sclerosis 784.51 dysarthria AND CPT E/M Service Code: 99201, 99202, 99203, 99204, 99205 (office-new patient), 99211,99212, 99213, 99214, 99215 (office-established patient), 99241, 99242, 99243, 99244, 99245 (outpatient consult), 99304, 99305, 99306, 99307, 99308, 99309, 99310 (nursing facility), 99324, 99325, 99326, 99327, 99328, 99334, 99335, 99336, 99337 (domiciliary), 99341, 99342, 99343, 99344, 99345, 99347, 99348, 99349, 99350 (home visit).
Numerator	Patients who were offered a referral at least once annually to a speech language pathologist for an augmentative/alternative communication evaluation. Reporting Instructions: • For all patients meeting denominator criteria, report the CPT Category II, **3762F**, *Patient is dysarthric* or **3763F**, *Patient is not dysarthric.* • When **3762F** is reported, also report **4552F,** *Patient offered referral to a speech language pathologist* 3762F- *Patient is dysarthric* 3763F-*Patient is not dysarthric* 4552F-*Patient offered referral to a speech language pathologist*
Denominator Exclusions	All patients with a diagnosis of amyotrophic lateral sclerosis who are dysarthric. • Documentation of a medical reason for not offering a referral to a speech language pathologist for an augmentative/alternative communication evaluation (eg patient is already using an augmentative communication device). Reporting Instructions: o For patient with appropriate exclusion criteria, report: **4552F-1P**

Measure #10: ALS End of Life Planning Assistance

Amyotrophic Lateral Sclerosis

Measure Description

Percentage of patients diagnosed with ALS who were offered at least once annually assistance in planning for end of life issues (eg advance directives, invasive ventilation, hospice).

Measure Components

Numerator Statement	Patients who were offered at least once annually assistance in planning for end of life issues (eg advance directives, invasive ventilation, or hospice).
Denominator Statement	All patients with a diagnosis of amyotrophic lateral sclerosis.
Denominator Exclusions	• Documentation of a medical reason for not offering at least once annually assistance in planning for end of life issues (eg patient in hospice and already in terminal phase)
Supporting Guideline & Other References	**The following clinical recommendation statements are quoted verbatim from the referenced clinical guidelines and represent the evidence base for the measure:** • Advance directives for palliative end-of-life care should be discussed early with the patient and carers, respecting the patient's social and cultural background.[1] • Offer assistance in formulating an advance care directive. (GPP)[2] • Review the patients' wishes regarding their care and advance directives regularly. (Level II)[3] • Re-discuss the patient's preferences for life-sustaining treatments every 6 months. (GPP)[2] • Initiate discussions on end-of-life issues whenever the patient asks or "opens the door" for end-of-life information and/or interventions. (GPP)[2]Treat pain in ALS following accepted guidelines. (GPP)[2] • Initiate early referral to hospice or home care teams well in advance of the terminal phase of ALS to facilitate the work of the hospice team. (GPP)[2] • Discuss options for respiratory support and end-of-life issues if the patient has dyspnea, other symptoms of hypoventilation or VC <50%. (GPP)[2] • Treat terminal dyspnea and/or pain with opioids alone or in combination with benzodiazepines if anxiety is present.(GPP)[2] [1] Andersen PM, Abrahams S, Borasio GD, et al. EFNS guidelines on the Clinical Management of Amyotrophic Lateral Sclerosis (MALS) - revised report of an EFNS task force. *Eur J Neurol* 2011;19(3) 360-375 (GPP=Good Practice Point) [2]Andersen PM, Borasio GD, Dengler R, et al. EFNS task force on management of amyotrophic lateral sclerosis: guidelines for diagnosing and clinical care of patients and relatives. *European J of Neurology* 2005;12:921-938 (GPP=Good Practice Point) [3]Heffernan C., Jenkinson C, Holmes T, et al. Management of respiration in MND/ALS patients: An evidence based review. *Amyotrophic Lateral Sclerosis* 2006; 7(1):5-15.

Measure Importance

Relationship to desired outcome

Referral to palliative services occurs varies considerably across different countries[1]. End of life discussions will improve patient decision making with respect to disease management [1-7] Offering assistance in formulating an advanced care directive can initiate this discussion and re-discussion of the patient's preferences for life-sustaining treatments every 6 months.[3,4,5] Pain in ALS should be treated following accepted guidelines.[8-11] Physical therapy will aid in treating spasticity and pain.[12] Discussion of respiratory support if the patient has dyspnea, other symptoms of hypoventilation or VC <50% will allow patient to choose intervention or hospice [12-13] Early referral to hospice or home care teams well in advance of the terminal phase of ALS will facilitate the work of the hospice team and improve patient transition to hospice.[15, 19] A medical social worker can help with financial issues. Medications for terminal dyspnea , pain and/or anxiety will improve quality of life [15-19]

References

[1] Bede P, Oliver D, Stodart J, van den Berg L, et al. Palliative care in amyotrophic lateral sclerosis: a review of current international guidelines and initiatives. *J Neurol Neurosurg Psychiatry* 2011; 82(4):413-8.
[2] Oliver D, Campbell C, Sykes N, et al. Decision-making for gastrostomy and ventilatory support for people with motor neurone disease: variations across UK hospices. *J Palliat Care* 2011; 27(3):198-201.
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Opportunity for

Palliative care should be adopted from the time of diagnosis.[1] Many patients are not

Improvement	adequately informed about advance directives and end of life decision making and many hospice workers are not familiar with ALS.[2,3,4] Management of the terminal phase of ALS is unsatisfactory in 33% - 61% of cases in Europe [5] and only 8% of palliative care units are involved from the time of diagnosis [6] The current system of palliative care in the USA is highly decentralized. [7] Between 60-88% of patients die in a medical facility in some countries and not at home, while over 58% of seriously ill ALS patients do not have hospice care[8, 9.,10]. Approaches to end of life care vary widely and are not standardized either in timing or content [1,11]

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IOM Domains of Health Care Quality Addressed	Effective Patient centered Timely
Exclusion Justification	A medical reason exclusion has been included for patients who are already in hospice and in the terminal phase.
Harmonization with Existing Measures	There exist two other measures that refer to advanced care planning. The American Geriatrics Society (2008) has a measure "Percentage of patients aged 65 years and older who have an advance care plan or surrogate decision maker documented in the medical record or documentation in the medical record that an advance care plan was discussed but the patient did not wish or was not able to name a surrogate decision maker or provide an advance care plan." However this measure is limited to those over the age of 65. The work group felt it was important to not exclude patients under 65 years old who have ALS from an end of life planning measure. In addition, the Institute for Clinical Systems Improvement (ICSI) (2009) has a measure that states the "Percentage of adult patients with the specified progressive, debilitating disease who have a palliative care plan* in chart." *A completed palliative care plan addresses all seven domains of care: physical aspects, cultural aspects, psychological aspects, social aspects, spiritual/ religious/existential aspects, ethical/legal

aspects, and care of the imminently dying patient. However, this measure does not reference specific end of life needs that are relevant for patients with ALS. This measure was also not developed by a medical specialty society and the methods used to develop the measure are unclear.

Measure Designation

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

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 –CM Diagnosis Codes: 335.20 amyotrophic lateral sclerosis AND CPT E/M Service Code: 99201, 99202, 99203, 99204, 99205 (office-new patient), 99211,99212, 99213, 99214, 99215 (office-established patient), 99241, 99242, 99243, 99244, 99245 (outpatient consult), 99304, 99305, 99306, 99307, 99308, 99309, 99310 (nursing facility), 99324, 99325, 99326, 99327, 99328, 99334, 99335, 99336, 99337 (domiciliary), 99341, 99342, 99343, 99344, 99345, 99347, 99348, 99349, 99350 (home visit)
Numerator	Patients who were offered at least once annually assistance in planning for end of life issues (eg advance directive, invasive ventilation, or hospice). Reporting Instructions: • For all patients meeting denominator criteria, report the CPT Category II, **4553F** *Patient offered assistance in planning for end of life issues* 4553F- *Patient offered assistance in planning for end of life issues*
Denominator Exclusions	All patients with a diagnosis of amyotrophic lateral sclerosis. • Documentation of a medical reason for not offering at least once annually assistance in planning for end of life issues at least once annually (eg patient in hospice and already in terminal phase)

Reporting Instructions:

- For patient with appropriate exclusion criteria, report: **4553F-1P**

Measure #11: ALS Falls Querying

Amyotrophic Lateral Sclerosis
For Quality Initiatives only

Measure Description

Percentage of visits for patients with a diagnosis of amyotrophic lateral sclerosis with patient queried about falls within the past 12 months.

Measure Components

Numerator Statement	Patient visits with patient queried about falls within the past 12 months.
Denominator Statement	All visits for patients with a diagnosis of amyotrophic lateral sclerosis.
Denominator Exclusions	• No exclusions applicable for this measure
Supporting Guideline & Other References	**The following clinical recommendation statements are quoted verbatim from the referenced clinical guidelines and represent the evidence base for the measure:** • An increased risk of falls is established among persons with diagnoses of stroke, dementia, and disorders of gait and balance (Level A) and probable among patients with Parkinson disease, peripheral neuropathy, lower extremity weakness or sensory loss, and substantial vision loss (Level B).[1] • As for screening measures that may predict or further assess fall risk, a history of recent falls is an established predictor of future falls (Level A).[1] [1]Thurman DJ, Stevens JA, Rao JK. Assessing patients in a neurology practice for risk of falls (an evidence-based review) : Report of the Quality Standards Subcommittee of the American Academy of Neurology *Neurology* 2008;70;473

Measure Importance

Relationship to desired outcome

Falls surveillance will lead to interventions to prevent falls and decrease fall related deaths in ALS patients. Falls are an independent predictor of adverse health outcomes.[1] Fall related deaths occur in 1.7% of ALS patients.[2] Several specific risk factors for falls have been identified, including muscle weakness, deficits in gait or balance, visual deficits, arthritis, impairments in activities of daily living, depression, and cognitive impairment.[3] In ALS clinical trials, falls are one of the most frequent adverse events in all patient groups. Falls are a major source of morbidity and mortality in ALS.[2]

References

[1]Laird RD, Studenski S, Perera S, Wallace D. Fall history is an independent predictor of adverse health outcomes and utilization in the elderly. *Am J Manag Care* 2001; 7(12):1133-8.
[2] Gil J, Funalot B, Verschueren A, et al. Causes of death amongst French patients with amyotrophic lateral sclerosis: a prospective study *Eur J Neurol.* 2008; 15(11):1245-51.
[3] Rubenstein LZ, Josephson KR. The epidemiology of falls and syncope. *Clin Geriatr Med* 2002; 18:141–158.

Opportunity for Improvement	Patients with neurologic or general conditions associated with an increased risk of falling should be asked about recent falls and further examined for the presence of specific neurologic deficits that predict falls, which include gait and balance disorders; deficits of lower extremity strength, sensation, and coordination; and cognitive impairments. If substantial risks of falls are identified, appropriate interventions that are described in other evidence-based guidelines may be considered.[1] There is an increased rate [Hazard ratio = 6.5 (95% CI 2.4-14.1)]of falls with head injury in ALS patients compared with a control population in the first year of ALS.[2] There is an increased prevalence of falls in ALS patients of 0.7 falls per patient per 6 months.[3] Falls surveillance will allow implementation of appropriate treatment interventions and home safety evaluations to prevent recurrence and potential adverse outcome. References [1]Thurman DJ, Stevens JA, Rao JK. Assessing patients in a neurology practice for risk of falls (an evidence-based review) : Report of the Quality Standards Subcommittee of the American Academy of Neurology *Neurology* 2008; 70;473 [2] Turner MR, Abisgold J, Yeates DGR, et al. Head and other physical trauma requiring hospitalization is not a significant risk factor in the development of ALS. *J Neurol Sci* 2010; 288 (1-2): 45-48. [3] Montes J, Cheng B, Diamond B, et al. The Timed Up and Go test: predicting falls in ALS. *Amyotroph Lateral Scler*. 2007; 8(5):292-5
IOM Domains of Health Care Quality Addressed	Safe Effective Patient centered
Exclusion Justification	No exclusions relevant for this measure.
Harmonization with Existing Measures	The American Geriatrics Society (AGS) measure was reviewed to assess the ability to harmonize this measure with their measure. It was determined that the age limitations on the AGS were not appropriate for ALS patients and that falls was enough of a safety issue that a separate falls and ALS measure was warranted.

Evidence from Quality Improvement Initiatives

Falls are one of the most common adverse effects associated with ALS. A falls question is included in almost every ALS trial. [1-2]

References
[1] Rosenfeld J, King RM, Jackson CE, et al. Creatine monohydrate in ALS: effects on strength, fatigue, respiratory status and ALSFRS. *Amyotroph Lateral Sc*ler 208 ; 9(5): 266-272.
[2]Aggarwal SP, Zinman L, Simpson E, et al and NEALS and Canadian Consortia. Safety and efficacy of lithium in combination with riluzole for treatment of amyotrophic lateral sclerosis: a randomized double-blind, placebo-controlled trial. *Lancet Neurology* 2010; 9:481-488.

Measure Designation

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

Level of Measurement	• Individual practitioner
Care setting	• Ambulatory Care
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

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 –CM Diagnosis Codes: 335.20 (amyotrophic lateral sclerosis) AND CPT E/M Service Code: 99201, 99202, 99203, 99204, 99205 (office-new patient), 99211,99212, 99213, 99214, 99215 (office-established patient), 99241, 99242, 99243, 99244, 99245 (outpatient consult), 99304, 99305, 99306, 99307, 99308, 99309, 99310 (nursing facility), 99324, 99325, 99326, 99327, 99328, 99334, 99335, 99336, 99337 (domiciliary), 99341, 99342, 99343, 99344, 99345, 99347, 99348, 99349, 99350 (home visit).
Numerator	Patient visits with patient queried about falls within the past 12 months. Reporting Instructions: • For all patients meeting the denominator criteria, report the CPT Category II, **6080F**, *Patient (or caregiver) queried about falls.*
Denominator Exclusions	All patients with a diagnosis of amyotrophic lateral sclerosis. Reporting Instructions: • No exclusions applicable for this measure. Do not report modifiers 1P, 2P, or 3P with 6080F.

Evidence Schemes

American Academy of Neurology

1. Miller RG, Jackson CE, Kasarskis EJ, et al. Practice Parameter update: The care of the patient with amyotrophic lateral sclerosis: Multidisciplinary care, symptom management, and cognitive/behavioral impairment (an evidence-based review): Report of the Quality Standards Subcommittee of the American Academy of Neurology *Neurology* 2009:73:1227-1233

2. Miller RG, Jackson CE, Kasarskis EJ, et al. Practice Parameter Update: The care of the patient with amyotrophic lateral sclerosis: Drug, nutritional, and respiratory therapies (an evidence-based review): Report of the Quality Standards Subcommittee of the American Academy of Neurology *Neurology* 2009; 73:1218-1226

3. Thurman DJ, Stevens JA, Rao JK. Assessing patients in a neurology practice for risk of falls (an evidence-based review) : Report of the Quality Standards Subcommittee of the American Academy of Neurology *Neurology* 2008;70;473

American Academy of Neurology (AAN) Rating Schemes

Suggested wording	Translation of evidence to recommendations	Rating of Therapeutic Article
(Note: Wording relevant to diagnostic, prognostic and screening questions are indicated in parenthesis.) **Conclusion:** A = Established as effective, ineffective or harmful (or established as useful/predictive or not useful/predictive) for the given condition in the specified population **Recommendation:** Should be done or, should not be done	Level A rating requires at least two consistent Class I studies*	**Class I:** Randomized, controlled clinical trial with masked or objective outcome assessment, in a representative population. Relevant baseline characteristics are presented and substantially equivalent among treatment groups or there is appropriate statistical adjustment for differences. The following are required: a) Concealed allocation b) primary outcome(s) clearly defined c) exclusion/inclusion criteria clearly defined d) adequate accounting for drop-outs (with at least 80% of enrolled subjects completing the study) and cross-overs with numbers sufficiently low to have minimal potential for bias
Conclusion: B = Probably effective, ineffective or harmful (or probably useful/predictive or not useful/predictive) for the given condition in the specified population **Recommendation:** Should be considered or, should not be considered	Level B rating requires at least one Class I study or two consistent Class II studies	**Class II:** Prospective matched group cohort study in a representative population with masked outcome assessment that meets b-d above OR a RCT in a representative population that lacks one criteria a-d.
Conclusion: C = Possibly effective, ineffective or harmful (or possibly useful/predictive or not useful/predictive) for the given condition in the specified population **Recommendation:** May be considered or, may not be considered	Level C rating requires at least one Class II study or two consistent Class III studies	**Class III:** All other controlled trials (including well-defined natural history controls or patients serving as own controls) in a representative population, where outcome is independently assessed, or independently derived by objective outcome measurement.**
Conclusion: U = Data inadequate or conflicting; given current knowledge, treatment (test, predictor) is unproven **Recommendation:** None	Studies not meeting criteria for Class I – Class III	**Class IV:** Studies not meeting Class I, II or III criteria including consensus, expert opinion or a case report.

*In exceptional cases, one convincing Class I study may suffice for an "A" recommendation if 1) all criteria are met, 2) the magnitude of effect is large (relative rate improved outcome > 5 and the lower limit of the confidence interval is > 2).

**Objective outcome measurement: an outcome measure that is unlikely to be affected by an observer's (patient, treating physician, investigator) expectation or bias (e.g., blood tests, administrative outcome data).

Rating of Diagnostic Article	Rating of Prognostic Article	Rating of Screening Article
Class I: A cohort study with prospective data collection of a broad spectrum of persons with the suspected condition, using an acceptable reference standard for case definition. The diagnostic test is objective or performed and **interpreted without knowledge of the patient's clinical status**. Study results allow calculation of measures of diagnostic accuracy.	**Class I:** A cohort study of a broad spectrum of persons at risk for developing the outcome (e.g. target disease, work status). The outcome is defined by an acceptable reference standard for case definition. The outcome is objective or measured by an observer who is masked to the presence of the risk factor. Study results allow calculation of measures of prognostic accuracy.	**Class I.** A statistical, population-based sample of patients studied at a uniform point in time (usually early) during the course of the condition. All patients undergo the intervention of interest. The outcome, if not objective, is determined in an evaluation that is masked to the patients' clinical presentations.
Class II: A case control study of a broad spectrum of persons with the condition established by an acceptable reference standard compared to a broad spectrum of controls or a cohort study where a broad spectrum of persons with the suspected condition where the data was collected retrospectively. The diagnostic test is objective or performed and **interpreted without knowledge of disease status**. Study results allow calculation of measures of diagnostic accuracy.	**Class II:** A case control study of a broad spectrum of persons with the condition compared to a broad spectrum of controls or a cohort study of a broad spectrum of persons at risk for the outcome (e.g. target disease, work status) where the data was collected retrospectively. The outcome is defined by an acceptable reference standard for case definition. The outcome is objective or measured by an observer who is masked to the presence of the risk factor. Study results allow calculation of measures of **prognostic accuracy**.	**Class II.** A statistical, non-referral-clinic-based sample of patients studied at a uniform point in time (usually early) during the course of the condition. Most patients undergo the intervention of interest. The outcome, if not objective, is determined in an evaluation that is masked to the patients' clinical presentations.
Class III: A case control study or cohort study where either persons with the condition or controls are of a narrow spectrum. The condition is established by an acceptable reference standard. The reference standard and diagnostic test are objective or performed and interpreted by different observers. Study results allow calculation of measures of diagnostic accuracy.	**Class III:** A case control study or a cohort study where either the persons with the condition or the controls are of a narrow spectrum where the data was collected retrospectively. The outcome is defined by an acceptable reference standard for case definition. The outcome is objective or measured by an observer who did not determine the presence of the risk factor. Study results allow calculation of measures of a prognostic accuracy.	**Class III.** A sample of patients studied during the course of the condition. Some patients undergo the intervention of interest. The outcome, if not objective, is determined in an evaluation by someone other than the treating physician.
Class IV: Studies not meeting Class I, II or III criteria including consensus, expert opinion or a case report.	**Class IV:** Studies not meeting Class I, II or III criteria including consensus, expert opinion or a case report.	**Class IV.** Studies not meeting Class I, II or III criteria including consensus, expert opinion or a case report.

Retrospective: a case control study. Prospective: a cohort survey. Objective: a measurement unlikely to be affected by expectation bias.

4. Andersen PM, Borasio GD, Dengler R, et al. EFNS task force on management of amyotrophic lateral sclerosis: guidelines for diagnosing and clinical care of patients and relatives. *European J of Neurology* 2005;12:921-938

Andersen PM, Abrahams S, Borasio GD, et al. EFNS guidelines on the Clinical Management of Amyotrophic Lateral Sclerosis (MALS) - revised report of an EFNS task force. *Eur J Neurol* 2011;19(3) 360-375

Level A - Established as useful/predictive or not useful/predictive for a diagnostic measure or established as effective, ineffective or harmful for a therapeutic intervention, and requires at least one convincing Class I study or at least two consistent, convincing Class II studies.
Level B - Established as useful/predictive or not useful/predictive for a diagnostic measure or established as effective, ineffective or harmful for a therapeutic intervention, and requires at least one convincing Class II study or overwhelming Class III evidence.
Level C - Established as useful/predictive or not useful/predictive for a diagnostic measure or established as effective, ineffective or harmful for a therapeutic intervention, and requires at least two Class III studies.

Level U - Not used
Good Clinical Practice (GCP) points - Recommended best practice based on the experience of the guideline development group. Usually based on Class IV evidence indicating large clinical uncertainty, such GCP points can be useful for health workers

Canadian Thoracic Society
1A – strong recommendation, high-quality of evidence
1B – strong recommendation, moderate quality of evidence
1C – strong recommendation, low-quality or very low quality evidence
2A – weak recommendation, high-quality evidence
2B – weak recommendation, moderate-quality evidence
2C – weak recommendation, low-quality or very low-quality evidence

5. Bott J, Blumenthal S, Buxton M, et al. Guidelines for the physiotherapy management of the adult, medical, spontaneously breathing patient. *Thorax* 2009; 64:1-51.

SIGN (Scottish Intercollegiate Guidelines Network):key to evidence statements and grades of recommendations
Levels of evidence
1++ High-quality meta-analyses, systematic reviews of randomised controlled trials (RCTs), or RCTs with a very low risk of bias
1+ Well-conducted meta-analyses, systematic reviews, or RCTs with a low risk of bias
12 Meta-analyses, systematic reviews, or RCTs with a high risk of bias
2++ High-quality systematic reviews of case–control or cohort studies
High-quality case–control or cohort studies with a very low risk of confounding or bias and a high probability that the relationship is causal
2+ Well-conducted case–control or cohort studies with a low risk of confounding or bias and a moderate probability that the relationship is causal
22 Case control or cohort studies with a high risk of confounding or bias and a significant risk that the relationship is not causal
3 Non-analytical studies (eg, case reports, case series)
4 Expert opinion

Grades of recommendations
A At least one meta-analysis, systematic review, or RCT rated as 1++, and directly applicable to the target population; or A body of evidence consisting principally of studies rated as 1+, directly applicable to the target population, and demonstrating overall consistency of results
B A body of evidence including studies rated as 2++, directly applicable to the target population, and demonstrating overall consistency of results; or Extrapolated evidence from studies rated as 1++ or 1+
C A body of evidence including studies rated as 2+, directly applicable to the target population and demonstrating overall consistency of results; or Extrapolated evidence from studies rated as 2++
D Evidence level 3 or 4; or Extrapolated evidence from studies rated as 2+
Good practice points *!* Recommended best practice based on the clinical experience of the guideline development group
Note: the grade of recommendation relates to the strength of the evidence on which the recommendation is based. It does not reflect the clinical importance of the recommendation.

6. National Institute for Health and Clinical Excellence (2010) Motor neurone disease: the use of non-invasive ventilation in the management of motor neurone disease. London: National Institute for Health and Clinical Excellence. Available from: www.nice.org.uk/guidance/CG105

Rating Scheme for Strength of the Evidence
Ia-Systematic review or meta-analysis of randomized controlled trials
Ib-At least one randomized controlled trial
IIa-At least one well-designed controlled stud without randomization
IIb-At least one well-designed quasi-experimental descriptive studies, such as a cohort study
III-Well-designed non-experimental descriptive studies, case-control studies, and case studies
IV-Expert committee reports, opinions and/or clinical experience of respected authorities

Rating Recommendations
A* Directly based on category I evidence (meta-analysis of randomized controlled trials (RCTs) or at least one RCT)
B* Directly based on category II evidence (at least one controlled study without randomization or at least one other quasi-experimental study) or extrapolated from category I evidence
C* Directly based on category III evidence (non-experimental descriptive studies) or extrapolated from category I or II evidence
D* Directly based on category III evidence (expert committee reports or opinions and/or clinical experience of respected authorities) or extrapolated from category I, II or III evidence
N Recommendation taken from NICE guideline or technology appraisal guidance

7. Heffernan C, Jenkinson C, Holmes T, et al. C. Management of respiration in MND/ALS patients: An evidence based review. *Amyotrophic Lateral Sclerosis* 2006; 7(1):5-15. (Systematic Review)

Grading of quality of evidence for the individual studies
1a Systematic review with homogeneity of RCTs – worrisome heterogeneity to be tagged with a "-".
1b Individual RCT with a narrow confidence interval
1c Quasi-experimental/RCT where all patients died before Rx became available but some now survive on it; or when some patients died before the Rx became available but none now die on it.
2a Systematic review with homogeneity of cohort studies
2b Individual cohort study (including low quality RCT; e.g., v80% follow-up)
2c 'Outcomes' Research (relates the health and clinical outcome of a cohort of patients with the same diagnosis to the care that they received)

3a Systematic review with homogeneity of case-control studies
3b Individual case-control study
3c Individual controlled cross-sectional study
4 Observational studies without controls (case series, before-and-after studies, prevalence cross-sectional studies) and poor quality cohort and case control studies
5 Expert opinion without explicit critical appraisal, or based on physiology, bench research or 'first principles' (i.e., the pathophysiological principles used to determine clinical practice)

Classification of recommendations for clinical management
I Strong recommendation for use in clinical management, arising from strong evidence obtained from systematic reviews with meta-analysis or randomised controlled trials
II Recommendation for clinical management that is based on experimental studies and/or observational studies
III Suggestion for clinical management that is based on expert opinion

8. Tripodoro VA, De Vito EL. Management of dyspnea in advanced motor neuron diseases. *Curr Opin Support Palliat Care* 2008;2(3):173-9. (Systematic Review)
No rating or evidence scheme available

9. Miller RG, Mitchell JD, Lyon M, More DH. Riluzole for amyotrophic lateral sclerosis (ALS)/motor neuron disease (MND) (Review). 2009 The Cochrane Library. 2:1-27.
No evidence or rating scheme available

Questions or Comments:

Please contact the American Academy of Neurology at quality@aan.com.