General

Title

Amyotrophic lateral sclerosis (ALS): percentage of patients diagnosed with ALS for whom a multidisciplinary care plan was developed, if not done previously, and the plan was updated at least once annually.

Source(s)


Measure Domain

Primary Measure Domain

Clinical Quality Measures: Process

Secondary Measure Domain

Does not apply to this measure

Brief Abstract

Description

This measure is used to assess the percentage of patients diagnosed with amyotrophic lateral sclerosis (ALS) for whom a multidisciplinary care plan was developed, if not done previously, and the plan was updated at least once annually.

Rationale

In specialized multidisciplinary clinics, patients with amyotrophic lateral sclerosis (ALS) receive comprehensive care from a neurologist, pulmonologist, gastroenterologist, psychiatrist, social worker, occupational therapist, speech language pathologist, respiratory therapist, specialized nurse case manager, physical therapist, dietitian, psychologist, dentist, and/or palliative care expert (EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis et al., 2012; Miller et al., 2009). 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 (Miller et al., 2009).

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.
Patient care and survival were examined for 97 patients attending specialized ALS clinics in Italy compared with 124 patients in neurology clinics (Chiò et al., 2006). 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 (Traynor et al., 2003). 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 (Traynor et al., 2003). Dutch patients in multidisciplinary ALS clinics (n=133) were compared with 75 patients receiving general care (Van den Berg et al., 2005). 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 (Zoccolella et al., 2007). 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.

The following clinical recommendation statements are quoted verbatim from the referenced clinical guidelines and represent the evidence base for the measure:

- Specialized multidisciplinary clinical referral should be considered for management of patients with ALS to optimize health care delivery and prolong survival and may be considered to enhance quality of life (Miller et al., 2009).
- Multidisciplinary care should be available for people affected by ALS as attendance at a multidisciplinary clinic improves care, and may extend survival (Andersen et al., 2005).
- 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 (Andersen et al., 2005).
- 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 (Heffernan et al., 2006).
- A palliative care approach should be incorporated into the care plan for patients and carers from the time of diagnosis (Tripodoro & De Vito, 2008).

Evidence for Rationale


Primary Health Components
Amyotrophic lateral sclerosis (ALS); multidisciplinary care plan

Denominator Description
All patients with a diagnosis of amyotrophic lateral sclerosis (ALS) (see the related "Denominator Inclusions/Exclusions" field)

Numerator Description
Patients for whom a multidisciplinary care plan was developed, if not done previously, and the plan was updated at least once annually (see the related "Numerator Inclusions/Exclusions" field)

Evidence Supporting the Measure
Type of Evidence Supporting the Criterion of Quality for the Measure
A clinical practice guideline or other peer-reviewed synthesis of the clinical research evidence

A formal consensus procedure, involving experts in relevant clinical, methodological, public health and organizational sciences

A systematic review of the clinical research literature (e.g., Cochrane Review)

One or more research studies published in a National Library of Medicine (NLM) indexed, peer-reviewed journal

Additional Information Supporting Need for the Measure

Importance of Topic

Prevalence and Incidence

- Amyotrophic lateral sclerosis (ALS), also known as Lou Gehrig’s disease, is a type of motor neuron disease that is a rapidly progressive and fatal neurological disease (National Institute of Neurological Disorders and Stroke [NINDS], 2013).
- Twenty thousand to 30,000 people in the United States (U.S.) have ALS (NINDS, 2013).
- Five thousand people are diagnosed with ALS in the U.S. annually (NINDS, 2013).
- ALS is one of the most common neuromuscular diseases worldwide (NINDS, 2013).
- In 90% to 95% of all ALS cases the disease occurs apparently at random with no clearly associated risk factors (NINDS, 2013).
- Five percent to 10% of all ALS cases are inherited (NINDS, 2013).
- Twenty percent of all familial cases result from a specific genetic defect that leads to mutation of the enzyme known as superoxide dismutase 1 (SOD1) (NINDS, 2013).
- No cure exists for ALS. Newer pharmacotherapy agents have been found to reduce the progression, but not halt the disease development (NINDS, 2013).
- 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 (ALS Association, 2012).
- Cognitive dysfunction is seen in 20% to 50%, while only 3% to 5% develop dementia that is usually of frontotemporal type (Strong et al., 2009). Consensus criteria for diagnosis have recently been reported (Strong et al., 2009).
- Death due to respiratory failure follows on average 2 to 4 years after onset, but a small group may survive for a decade or more (Haverkamp, Appel, & Appel, 1995).
- The mean age of onset is 47 to 52 years in familial cases (FALS) and 58 to 63 years in sporadic (SALS) cases (Bobowick & Brody, 1973).
- 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 (Armon, 2007) with male sex, increasing age and hereditary disposition being the main risk factors (Heffernan et al., 2006).

Mortality and Morbidity

- Most patients with ALS die within 2 to 5 years of onset (Lechtzin et al., 2002). Only 10% of ALS patients survive for 10 years or more (Miller et al., “Drug, nutritional,” 2009).
- Treatment of respiratory insufficiency improves survival, quality of life and respiratory symptoms (Lechtzin et al., 2002; Miller et al., “Drug, nutritional,” 2009). The diagnosis and management of respiratory insufficiency is critical because most deaths from ALS are due to respiratory failure (Lechtzin et al., 2002; Miller et al., “Drug, nutritional,” 2009; EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis et al., 2012; Laird et al., 2001).
- 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 (Gil et al., 2008). Fall related deaths occur in 1.7% of ALS patients (Rubenstein & Josephson, 2002). 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 (Ringholz et al., 2005).
- 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 (Laird et al., 2001). More severe impairment occurs in a subset of patients with ALS and has features consistent with frontotemporal dementia (FTD) (Phukan, Pender, & Hardiman, 2007; Gordon et al., 2007). Recent studies have demonstrated the feasibility of screening patients in a busy specialized ALS clinic (Flaherty-Craig et al., 2009; Woolley & Katz, 2011), 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 (Elamin et al., 2011), 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 motor neuron disease (MND).
• Pseudobulbar affect (PBA), excessive laughing or crying, or involuntary emotional expression disorder affects 20% to 50% of patients with ALS, especially in pseudobulbar palsy (McCullagh et al., 1999). Patients are embarrassed and isolated by these symptoms, which in turn greatly diminishes the patients’ quality of life.

• Salivary, 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 (Laird et al., 2001; Miller et al., "Multi-disciplinary," 2009)

• 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 (Bensimon, Lacomblez, & Meininger, 1994). Asthenia occurred in 18% of patients taking riluzole vs. 12% of patients taking placebo in a larger study (Lacomblez et al., 1996).

• The prevalence of depression in ALS ranges from 0% to 44%, although systematic studies suggest 10% in advanced ALS (Laird et al., 2001; Wicks et al., 2007). Depression shortens survival and lowers quality of life for patients with ALS (Phukan, Pender, & Hardiman, 2007). There is consensus among experts that depression should be treated in patients with ALS (Laird et al., 2001); 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 (Hetta & Jansson, 1997). 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 less than 18.5 kg/m² (Marin et al., 2011; Lehmbröck et al., 2012; Spataro et al., 2011; Desport et al., 1999; Vaisman et al., 2009; Dupuis et al., 2008).

• 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 (Ball, Beukelman, & Pattee, "Communication," 2004; Ball, Beukelman, & Pattee, "Acceptance," 2004; Mathy, Yorkston, & Gutmann, 2000; Beukelman, Fager, & Nordness, 2011).

• End of life discussions will improve patient decision making with respect to disease management (NINDS, 2013; ALS Association, 2012; Strong et al., 2009; Haverkamp, Appel, & Appel, 1995; Bobowick & Brody, 1973; Heffernan et al., 2006). Pain in ALS should be treated following accepted guidelines (Oliver et al., 2011; Albert et al., 1999, Mitsumoto et al., 2005; Nolan et al., 2008; Albert et al., 2005; Albert et al., 2009).

**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 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 (Dubinsky, Chen, & Lai, 2006).

**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 (Chiò et al., 2005).

• 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 (Thommen & Sambataro, 2001).

**Cost**

• 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 U.S. 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 (Ginsberg & Lowe, 2002).

Disparities
- All races and ethnic backgrounds are affected by ALS (NINDS, 2013).
- ALS most common in individuals 40 to 60 years old, but younger and older people can develop the disease (NINDS, 2013).
- 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 (ALS Association, 2012).

Opportunity for Improvement
- Treatments for ALS are underutilized even in specialized clinics (Bradley et al., 2004; Miller et al., "Outcomes," 2009). Studies suggest that even in tertiary care centers, there are varying degrees of adherence to the evidence-based American Academy of Neurology (AAN) practice parameters (Bradley et al., 2004; Miller et al., "Outcomes," 2009).
- Recent studies show that there is a much higher utilization rate of evidence-based treatments in multidisciplinary clinics than in community-based care (Miller et al., "Outcomes," 2009). 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 (Bradley et al., 2004; Miller et al., "Outcomes," 2009).
- 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.

Evidence for Additional Information Supporting Need for the Measure


**Extent of Measure Testing**

This measure is being made available without any prior testing. The American Academy of Neurology (AAN) recognizes the importance of testing of all its measures and encourages testing of the amyotrophic lateral sclerosis (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.
Evidence for Extent of Measure Testing

State of Use of the Measure

State of Use
Current routine use

Current Use
not defined yet

Application of the Measure in its Current Use

Measurement Setting
Ambulatory/Office-based Care
Home Care
Hospital Outpatient
Skilled Nursing Facilities/Nursing Homes

Professionals Involved in Delivery of Health Services
not defined yet

Least Aggregated Level of Services Delivery Addressed
Individual Clinicians or Public Health Professionals

Statement of Acceptable Minimum Sample Size
Does not apply to this measure

Target Population Age
Unspecified

Target Population Gender
Either male or female
National Strategy for Quality Improvement in Health Care

National Quality Strategy Aim

Better Care

National Quality Strategy Priority

Person- and Family-centered Care
Prevention and Treatment of Leading Causes of Mortality

Institute of Medicine (IOM) National Health Care Quality Report Categories

IOM Care Need

Living with Illness

IOM Domain

Effectiveness
Patient-centeredness

Data Collection for the Measure

Case Finding Period

Unspecified

Denominator Sampling Frame

Patients associated with provider

Denominator (Index) Event or Characteristic

Clinical Condition

Denominator Time Window

not defined yet

Denominator Inclusions/Exclusions

Inclusions
All patients with a diagnosis of amyotrophic lateral sclerosis (ALS)

Exclusions
Documentation of a system reason for not developing and updating annually a multidisciplinary care plan (e.g., patient has no insurance to cover a multidisciplinary plan)

Exclusions/Exceptions
not defined yet

Numerator Inclusions/Exclusions

Inclusions
Patients for whom a multidisciplinary care plan* was developed, if not done previously, and the plan was updated at least once annually

*Multidisciplinary 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.

Exclusions
Unspecified

Numerator Search Strategy
Fixed time period or point in time

Data Source
Administrative clinical data
Electronic health/medical record
Paper medical record

Type of Health State
Does not apply to this measure

Instruments Used and/or Associated with the Measure
Unspecified

Computation of the Measure

Measure Specifies Disaggregation
Does not apply to this measure

Scoring
Interpretation of Score
Desired value is a higher score

Allowance for Patient or Population Factors
not defined yet

Standard of Comparison
not defined yet

Identifying Information

Original Title
Measure #1: ALS multidisciplinary care plan developed or updated.

Measure Collection Name
Amyotrophic Lateral Sclerosis Performance Measurement Set

Submitter
American Academy of Neurology - Medical Specialty Society

Developer
American Academy of Neurology - Medical Specialty Society

Funding Source(s)
Unspecified

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Financial Disclosures/Other Potential Conflicts of Interest

Unspecified

Adaptation

This measure was not adapted from another source.

Date of Most Current Version in NQMC

2012 Jul

Measure Maintenance

Unspecified

Date of Next Anticipated Revision

Unspecified

Measure Status

This is the current release of the measure.

Measure Availability

Source available from the American Academy of Neurology (AAN) Web site.

For more information, contact AAN at 201 Chicago Avenue, Minneapolis, MN 55415; Phone: 800-879-1960; Fax: 612-454-2746; Web site: www.aan.com.

NQMC Status