



Ph 920-674-6255 • Fax 920-674-5288

**DETERMINING TERMINAL STATUS
AMYOTROPHIC LATERAL SCLEROSIS [ALS]**

Patient Name: _____

MR#: _____

DOB: _____

Admission Date: _____

Attending Physician: _____

Patients will be considered to be in the terminal stage of the individuals' prognosis (**life expectancy of six months or less**) if the terminal illness runs its normal course.

General Considerations:

1. ALS tends to progress in a linear fashion over time. Thus the *overall* rate of decline in each patient is fairly constant and predictable, unlike many other non-cancer diseases.
2. However, no *single* variable deteriorates at a uniform rate in all patients. Therefore, multiple clinical parameters are required to judge the progression of ALS.
3. Although ALS usually presents in a localized anatomical area, the location of initial presentation does not correlate with survival time. By the time patients become end-stage, muscle denervation has become widespread, affecting all areas of the body, and initial predominance patterns do not persist.
4. Progression of disease differs markedly from patient to patient. Some patients decline rapidly and die quickly; others progress more slowly. For this reason, the history of progression in individual patients is important to obtain to predict prognosis.
5. In end-stage ALS, two factors are critical in determining prognosis; ability to breathe, and to a lesser extent ability to swallow. The former can be managed by artificial ventilation, and the latter by gastrostomy or other artificial feeding, unless the patient has recurrent aspiration pneumonia. While not necessarily a contraindication to Hospice Care, the decision to institute either artificial ventilation or artificial feeding will significantly alter six-month prognosis.

Examination by a neurologist within three months of assessment for hospice is advised, both to confirm the diagnosis and to assist with prognosis.

Criteria:

Patients will be considered to be in the terminal stage of ALS (life expectancy of six months or less if they meet the following criteria. (Should fulfill 1, 2, or 3.)

1. Critically impaired breathing capacity as demonstrated by all the following characteristics occurring within the 12 months preceding initial hospice certification:
 - Vital capacity (VC) less than 30% of normal (if available).
 - Dyspnea at rest.
 - Requiring supplemental oxygen at rest.
 - Patient declines artificial ventilation; external ventilation used for comfort measures only.

Criteria:

Patient should demonstrate both rapid progression of ALS and critical nutritional impairment.

2. Rapid progression of ALS as demonstrated by all the following characteristics occurring within the 12 months preceding initial hospice certification.
 - Progression from independent ambulation to wheelchair to bedbound status.
 - Progression from normal to barely intelligible or unintelligible speech.
 - Progression from normal to pureed diet.
 - Progression from independence in most or all activities of daily living (ADLs) to needing major assistance by caretaker in all ADLs.
3. Critical nutritional impairment as demonstrated by all of the following characteristics occurring within the 12 months preceding initial hospice certification.

- Oral intake of nutrients and fluids insufficient to sustain life.
- Continuing weight loss.
- Dehydration or hypovolemia.
- Absence of artificial feeding methods sufficient to sustain life, but not for relieving hunger.

Criteria:

Patient should demonstrate both rapid progression of ALS and life-threatening complications.

4. Life threatening complications as demonstrated by one of the following characteristics occurring within the 12 months preceding initial hospice certification.

- Recurrent aspiration pneumonia (with or without tube feedings).
- Upper urinary tract infection, e.g., pyelonephritis.
- Sepsis.
- Recurrent fever after antibiotic therapy.
- Stage 3 or 4 decubitus ulcer(s).

Clinical Status

Progression of disease as documented by worsening clinical status, symptoms, signs and laboratory results.

- Recurrent or intractable infections such as pneumonia, sepsis or upper urinary tract.
- Weight loss not due to reversible causes such as depression or use of diuretics.
- Decreasing anthropomorphic measurements, not due to reversible causes such as depression or use of diuretics.
- Decreasing serum albumin or cholesterol.
- Dysphagia leading to recurrent aspiration and/or inadequate oral intake documented by decreasing food portion consumption.

Symptoms

- Dyspnea with increasing respiratory rate.
- Cough, intractable.
- Nausea/vomiting poorly responsive to treatment.
- Diarrhea, intractable.
- Pain requiring increasing doses of major analgesics more than briefly.

Signs

- Decline in systolic blood pressure to below 90 or progressive postural hypotension.
- Ascites.
- Venous, arterial or lymphatic obstruction due to local progression or metastatic disease.
- Edema.
- Pleural/pericardial effusion.
- Weakness.
- Change in level of consciousness.

Laboratory (When available. Lab testing is not required to establish hospice eligibility.)

- Increasing pCO₂ or decreasing pO₂ or decreasing SaO₂.
- Increasing calcium, creatinine or liver function studies.
- Increasing tumor markers (e.g., CEA, PSA).
- Progressively decreasing or increasing serum sodium or increasing serum potassium.
- Decline in Karnofsky Performance Status (KPS) or Palliative Performance Score (PPS) from \leq 70% due to progression of disease.
- Increasing emergency room visits, hospitalizations, or physician's visits related to hospice primary diagnosis.
- Progressive decline in Functional Assessment Staging (FAST) for dementia (from \geq 7A on the FAST).

- Progression to dependence on assistance with additional activities of daily living.
- Progressive stage 3-4 pressure ulcers in spite of optimal care.
- Physiologic impairment of functional status as demonstrated by:
 Karnofsky Performance Status (KPS) or Palliative Performance Score (PPS)
 from $\leq 70\%$. Note that two of the disease specific guidelines (HIV Disease,
 Stroke and Coma) establish lower qualifying KPS or PPS.

Dependence on assistance for two or more activities of daily living (ADLs).

- Feeding
- Ambulation
- Continence
- Transfer
- Bathing
- Dressing

Co-morbidities

- Chronic obstructive pulmonary disease.
- Congestive heart failure.
- Ischemic heart disease.
- Diabetes mellitus.
- Neurologic disease (CVA, ALS, MS, Parkinson's).
- Renal failure.
- Liver disease.
- Neoplasia.
- Acquired immune deficiency syndrome.
- Dementia.

Other comments or supporting documentation:

Please read and/or update this LMRP. If you agree with the documentation defining the terminal status of this patient, please sign below and FAX back.

Signature _____ **Date** _____