

Hospice -- Determining Terminal Status

Amyotrophic Lateral Sclerosis (ALS)

SPECIFIC INDICATIONS: A patient will be considered to have a life expectancy of six months or less if he/she meets the non-disease specific decline in clinical status guidelines described in Part I. Alternatively, the baseline non-disease specific guidelines described in Part II plus the applicable disease specific guidelines listed will establish the necessary expectancy.

Part I. Decline in clinical status guidelines

Patients will be considered to have a life expectancy of six months or less if there is documented evidence of decline in clinical status based on the guidelines listed below. Since determination of decline presumes assessment of the patient's status over time, it is essential that both baseline and follow-up determinations be reported where appropriate. Baseline data may be established on admission to hospice or by using existing information from records. Other clinical variables not on this list may support a six-month or less life expectancy. These should be documented in the clinical record.

These changes in clinical variables apply to patients whose decline is not considered to be reversible. They are listed in order of their likelihood to predict poor survival, the most predictive first and the least predictive last. No specific number of variables must be met, but fewer of those listed first (more predictive) and more of those listed last (least predictive) would be expected to predict longevity of six months or less.

Part I. Decline in clinical status guidelines

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

A. Clinical Status

- 1) Recurrent or intractable infections such as pneumonia, sepsis or upper urinary tract.
- 2) Progressive inanition as documented by:
 - a) Weight loss not due to reversible causes such as depression or use of diuretics
 - b) Decreasing anthropomorphic measurements (mid-arm circumference, abdominal girth), not due to reversible causes such as depression or use of diuretics
 - c) Decreasing serum albumin or cholesterol
- 3) Dysphagia leading to recurrent aspiration and/or inadequate oral intake documented by decreasing food portion consumption.

B. Symptoms

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

C. Signs

- 1) Decline in systolic blood pressure to below 90 or progressive postural hypotension
- 2) Ascites
- 3) Venous, arterial or lymphatic obstruction due to local progression or metastatic disease
- 4) Edema
- 5) Pleural / pericardial effusion
- 6) Weakness
- 7) Change in level of consciousness

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

- A. Increasing pCO₂ or decreasing pO₂ or decreasing SaO₂
 - B. Increasing calcium, creatinine or liver function studies
 - C. Increasing tumor markers (e.g. CEA, PSA)
 - D. Progressively decreasing or increasing serum sodium or increasing serum potassium
2. Decline in Karnofsky Performance Status (KPS) or Palliative Performance Score (PPS) from <70% due to progression of disease.
 3. Increasing emergency room visits, hospitalizations, or physician's visits related to hospice primary diagnosis
 4. Progressive decline in Functional Assessment Staging (FAST) for dementia (from 7A on the FAST)
 5. Progression to dependence on assistance with additional activities of daily living (See Part II, Section 2)
 6. Progressive stage 3-4 pressure ulcers in spite of optimal care

Part II. Non-disease specific baseline guidelines

(both of these should be met)

1. Physiologic impairment of functional status as demonstrated by: Karnofsky Performance Status (KPS) or Palliative Performance Score (PPS) <70%.

Note: two of the disease specific guidelines (HIV Disease, Stroke and Coma) establish a lower qualifying KPS or PPS.

2. Dependence on assistance for two or more activities of daily living (ADLs)
 - A. Feeding
 - B. Ambulation
 - C. Continence
 - D. Transfer
 - E. Bathing
 - F. Dressing

Amyotrophic Lateral Sclerosis

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 the rate of progression in individual patients is important to obtain to predict prognosis.
5. In end-state 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.
6. 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. Patient should demonstrate critically impaired breathing capacity.
 - a. Critically impaired breathing capacity as demonstrated by all the following characteristics occurring within the 12 months preceding initial hospice certification:
 - i. Vital capacity (VC) less than 30% of normal (if available);
 - ii. Dyspnea at rest;
 - iii. Patient declines mechanical ventilation; external ventilation used for comfort measures only.

Documentation Requirements

2. Patient should demonstrate both rapid progression of ALS and critical nutritional impairment.
 - a. Rapid progression of ALS as demonstrated by all the following characteristics occurring within the 12 months preceding initial hospice certification:
 - i. Progression from independent ambulation to wheelchair to bed bound status;
 - ii. Progression from normal to barely intelligible or unintelligible speech;
 - iii. Progression from normal to pureed diet;
 - iv. Progression from independence in most or all activities of daily living (ADLs) to needing major assistance by caretaker in all ADLs.
 - b. Critical nutritional impairment as demonstrated by all the following characteristics occurring within the 12 months preceding initial hospice certification:
 - i. Oral intake of nutrients and fluids insufficient to sustain life;
 - ii. Continuing weight loss;
 - iii. Dehydration or hypovolemia;
 - iii. Absence of artificial feeding methods, sufficient to sustain life, but not for relieving hunger.
3. Patient should demonstrate both rapid progression of ALS and life-threatening complications.
 - a. Rapid progression of ALS, see 2.a above.
 - b. Life-threatening complications as demonstrated by one of the following characteristics occurring within the 12 months preceding initial hospice certification:
 - i. Recurrent aspiration pneumonia (with or without tube feedings);
 - ii. Upper urinary tract infection, e.g., pyelonephritis;
 - iii. Sepsis;
 - iv. Recurrent fever after antibiotic therapy;
 - v. Stage 3 or 4 decubitus ulcer(s).

The baseline guidelines do not independently qualify a patient for hospice coverage.

Note: The word “should” in the disease specific guidelines means that on medical review the guideline so identified will be given great weight in making a coverage determination. It does not mean, however, that meeting the guideline is obligatory.

Part III. Co-morbidities

Although not the primary hospice diagnosis, the presence of disease such as the following, the severity of which is likely to contribute to a life expectancy of six months or less, should be considered in determining hospice eligibility.

- A. Chronic obstructive pulmonary disease
- B. Congestive heart failure
- C. Ischemic heart disease
- D. Diabetes mellitus
- E. Neurologic disease (CVA, ALS, MS, Parkinson’s)
- F. Renal failure
- G. Liver Disease
- H. Neoplasia
- I. Acquired immune deficiency syndrome
- J. Dementia