



MANAGE ALS FROM THE BEGINNING: CARE MAKES A DIFFERENCE

Although advances in understanding of the pathophysiology of ALS have stimulated the development of new drug therapies, the mainstay of treatment of ALS patients remains symptomatic management. This practice parameter summary comprises the first recommendation for the management of ALS that are based on a prescribed review and analysis of peer-reviewed literature. It was developed to improve the care and the quality of life of people with ALS by providing a rational basis for managing the disease.

Visit www.aan.com/professionals/practice/index.cfm to view the entire guideline.

Managing the Symptoms

In managing symptoms to improve the quality of life of the patient, family, and health care provider, evidence supports the following:

Nutrition

Consider PEG (percutaneous endoscopic gastrostomy) soon after symptom onset for patients who have symptomatic dysphagia (B). For optimal safety and efficacy, offer and implement PEG when the patient's vital capacity is above 50% of predicted (B). (See treatment algorithm on back).

Respiration

Deciding when to initiate noninvasive mechanical ventilation is critical because of the risk of either sudden death or ventilator dependence. Be vigilant for symptoms indicating hypoventilation (B). For best results in prolonging patient survival, initiate therapy before the patient's vital capacity falls below 50% of predicted (B). Respect the patient's right to refuse or withdraw any treatment, including mechanical ventilation. When withdrawing ventilation, use adequate opiates and anxiolytics to relieve dyspnea and anxiety (B). (See treatment algorithm on back).

Sialorrhea

Treat sialorrhea with glycopyrrolate, benztropine, transdermal hyoscine, atropine, trihexyphenidyl hydrochloride, or amitriptyline (C).

Pseudobulbar affect (emotional lability)

Treat pseudobulbar affect with amitriptyline or consider fluvoxamine as an alternate choice (C).

Palliative Care

As ALS progresses, the goal of patient care changes from maximizing function to providing effective and compassionate palliative care. Two of the most prevalent and unpleasant symptoms in the terminal phase are dyspnea and anxiety. Dyspnea can be treated with opioids alone or with supplemental oxygen (B) or possibly with chlorpromazine or acupuncture (C). Anxiety can be treated with anxiolytics such as lorazepam or diazepam, opioids, or chlorpromazine. Hospice care should be considered.

Breaking the News

- Give the diagnosis in person and never by telephone (B).
- Discuss the implications of the diagnosis, respecting the patient's cultural and social background by asking if the patient wishes to receive the information or have it communicated to a family member (B).
- Provide written reference materials about the disease and about support and advocacy organizations (B), as well as a letter or audio tape summarizing what you have discussed (B).

How Should a Physician Break Bad News to a Patient

(adapted from Ptacek, Eberhardt 1996)

Location

- Quiet, comfortable, and private

Structure

- In person, face-to-face
- Convenient time
- Enough time to ensure no rushing or interruptions
- Make eye contact and sit close to patient

Participants

- Have patient's support network available

What is said

- Find out what the patient already knows about the condition
- Ascertain how much the patient wants to know about the disease
- Give a warning comment that bad news is coming
- Acknowledge and explore patient's reaction and allow for emotional expression
- Summarize the discussion verbally, in writing, and/or audiotape
- Allow for questions

Reassurance

- Explain whether the complications of the disease are treatable
- Indicate that every attempt will be made to maintain function and patient's decisions will be respected
- Reassure that care for the patient will continue
- Discuss opportunities to participate in research treatment protocols
- Acknowledge willingness to get a second opinion if the patient wishes

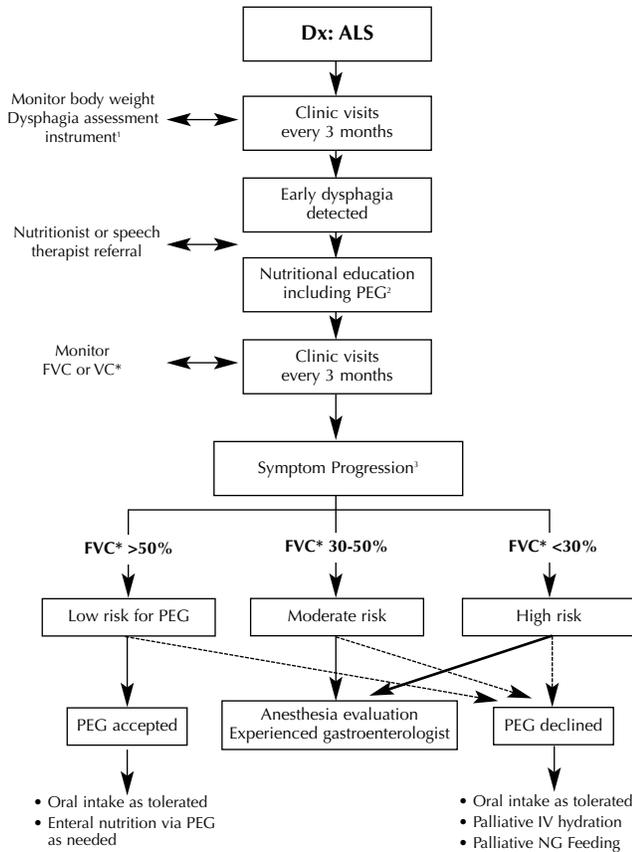
How it is said

- Emotional manner: warmth, caring, empathy, respect
- Give news at person's pace; allow patient to dictate what he or she is told

Language

- Simple words, yet direct; no euphemisms or medical jargon

ALGORITHM FOR NUTRITION MANAGEMENT



¹Rule out contraindications

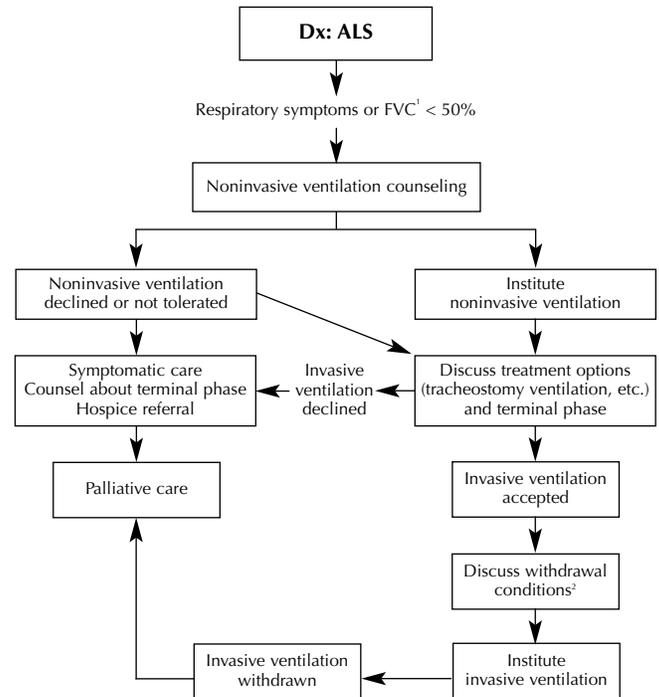
²Prolonged mealtime, ending meal prematurely because of fatigue, accelerated weight loss due to poor caloric intake, family concern about feeding difficulties

^{*}Forced vital capacity (FVC) or vital capacity (VC) can be used. VC may be more accurate in patients with bulbar dysfunction

³For example, Colorado Dysphagia Disability Inventory, bulbar questions in the ALS Functional Rating Scale, or other instrument

Dx = diagnosis PEG = percutaneous endoscopic gastrostomy

ALGORITHM FOR RESPIRATORY MANAGEMENT



¹Forced vital capacity (FVC) or vital capacity (VC) can be used. VC may be more accurate in patients with bulbar dysfunction

²Agreement needed for conditions of withdrawal prior to or concurrent with instituting invasive ventilation (e.g. locked in state, coma, etc.)

Dx = diagnosis

Classification of management recommendations

Classification	Definition
Standard	A principle for patient management that reflects a high degree of certainty based on class I evidence, or very strong evidence from class II studies when circumstances preclude randomized trials (A)
Guideline	Recommendations for patient management reflecting moderate clinical certainty (usually class II evidence or strong consensus of class III evidence) (B)
Option	A strategy for patient management for which the evidence (class III) is inconclusive or when there is some conflicting evidence or opinion (C)

This is an evidence-based educational service of the American Academy of Neurology. It is designed to provide members with evidence-based guideline recommendations to assist with decision-making in patient care. It is based on an assessment of current scientific and clinical information, and is not intended to exclude any reasonable alternative methodologies. The AAN recognizes that specific patient care decisions are the prerogative of the patient and the physician caring for the patient, based on the circumstances involved. Physicians are encouraged to carefully review the full AAN guidelines so they understand all recommendations associated with care of these patients.

Copies of this summary are available at www.aan.com/professionals/practice/index.cfm or through AAN Member Services at (800) 879-1960.



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