AAN Summary of Evidence-based Guideline for CLINICIANS

THE CARE OF THE PATIENT WITH AMYOTROPHIC LATERAL SCLEROSIS: DRUG, NUTRITIONAL, AND RESPIRATORY THERAPIES

This is a summary of the American Academy of Neurology (AAN) guideline regarding management and care of the patient with amyotrophic lateral sclerosis (ALS). Recommendations are presented for drug, nutritional, and respiratory therapies.

Please refer to the full guideline at www.aan.com for more information, including the AAN's definitions of the levels of recommendations and classifications of evidence.

DRUG THERAPIES	
What is the effect of riluzole on slowing the disease process or prolonging survival in ALS?	
Strong evidence	Riluzole should be offered to slow disease progression in patients with ALS (**Level A**).
Does lithium carbonate prolong survival or slow disease progression in ALS?	
Insufficient evidence	There are insufficient data at this time to support or refute treatment with lithium carbonate in patients with ALS (**Level U**).
NUTRITIONAL THERAPIES	
What is the effect of enteral nutrition administered via percutaneous endoscopic gastrostomy (PEG) on weight stability?	
Good evidence	In patients with ALS with impaired oral food intake, enteral nutrition via PEG should be considered to stabilize body weight (**Level B**).
When is PEG indicated in ALS?	
Insufficient evidence	There are insufficient data to support or refute specific timing of PEG insertion in patients with ALS (**Level U**).
What is the efficacy of nutritional support via PEG in prolonging survival?	
Good evidence	PEG should be considered for prolonging survival in patients with ALS (**Level B**).
What is the effect of enteral nutrition delivered via PEG on quality of life (QOL)?	
Insufficient evidence	There are insufficient data to support or refute PEG for improving QOL in patients with ALS (**Level U**).
What is the efficacy of vitamin and nutritional supplements on prolonging survival or QOL?	
Strong evidence	Creatine, in doses of 5 to 10 g daily, should not be given as treatment for ALS because it is not effective in slowing disease progression (**Level A**).
Good evidence	High-dose vitamin E should not be considered as treatment for ALS (**Level B**).
Insufficient evidence	The equivocal evidence regarding low-dose vitamin E permits no recommendation (**Level U**).
RESPIRATORY THERAPIES	
What are the optimal pulmonary tests to detect respiratory insufficiency?	
Weak evidence	Nocturnal oximetry may be considered to detect hypoventilation (regardless of the forced vital capacity [FVC]) (**Level C**).
	Supine FVC and maximal inspiratory pressure (MIP) may be considered useful in routine respiratory monitoring, in addition to the erect FVC (**Level C**).
	Sniff nasal pressure (SNP) may be considered to detect hypercapnia and nocturnal hypoxemia (**Level C**).
Does noninvasive ventilation (NIV) improve respiratory function or increase survival?	
Good evidence	NIV should be considered to treat respiratory insufficiency in ALS, both to lengthen survival and to slow the rate of FVC decline (**Level B**).
How do invasive ventilation and NIV affect QOL?	
Weak evidence	NIV may be considered to enhance QOL in patients with ALS who have respiratory insufficiency (**Level C**).
	Tracheostomy invasive ventilation (TIV) may be considered to preserve QOL in patients with ALS who want long-term ventilatory support. (**Level C**).
What factors influence acceptance of invasive ventilation and NIV?	
Weak evidence	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**).
What is the efficacy of targeted respiratory interventions for clearing secretions?	
Weak evidence	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**).

Insufficient evidence	There are insufficient data to support or refute high-frequency chest wall oscillation (HFCWO) for clearing airway secretions in patients with ALS (**Level U**).
Clinical context*	Medications with mucolytics, a B-receptor antagonist, nebulized saline, or an anticholinergic bronchodilator are widely used; however, no controlled studies exist in ALS.

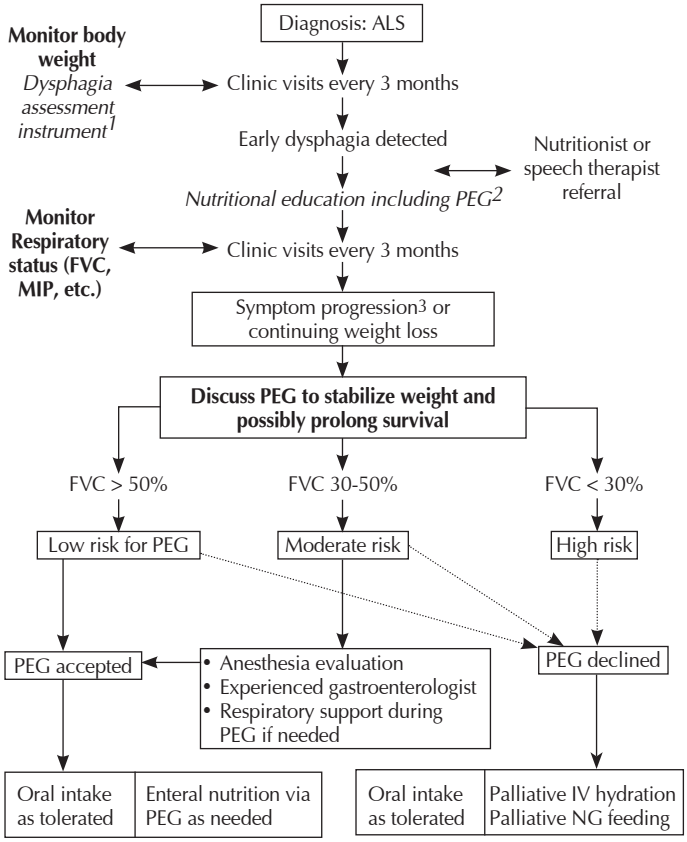
CLINICAL CONTEXT*

The ALS patient CARE database was developed with the hope of standardizing new and effective therapies for patients with ALS and tracking outcomes to raise the standard of care. Data obtained from the ALS CARE program have shown that the underutilization of many therapies has persisted in the years since the previous practice parameter, though there have been gains. These findings suggest that an evidence-based practice parameter may over time become more widely accepted and change practice. However, the persistent underutilization of therapies that improve survival and QOL poses a challenge for ALS clinicians to continue to raise the standard of care for patients with ALS.

*Clinical context slightly abridged. See the published guideline for the complete text.

The following figures present approaches to nutritional and respiratory management in patients with ALS.

Figure 1. Nutrition management algorithm



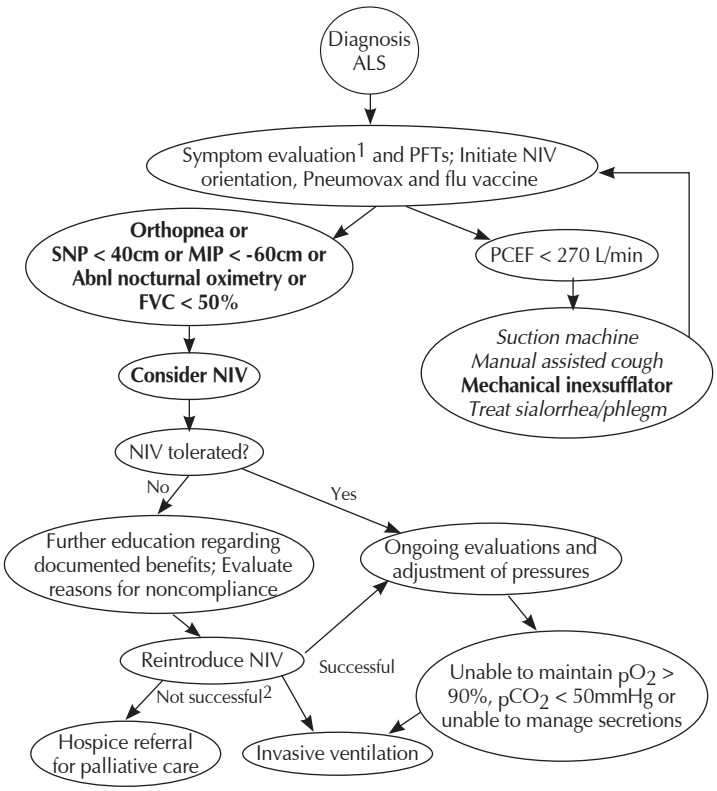
1e.g., bulbar questions in the Amyotrophic Lateral Sclerosis Functional Rating Scale, or other instrument.

2Percutaneous endoscopic gastrostomy: rule out contraindications.

3Prolonged meal time; ending meal prematurely because of fatigue; accelerated weight loss due to poor caloric intake; family concern about feeding difficulties.

Text in bold = evidence-based
Text in italics = consensus-based

Figure 2. Respiratory management algorithm



1Symptoms suggestive of nocturnal hypoventilation:
- *Frequent arousals*
- *Morning headaches*
- *Excessive daytime sleepiness*
- *Vivid dreams*

2If NIV is not tolerated or accepted in the setting of advancing respiratory compromise, consider invasive ventilation or referral to hospice.

PFT = pulmonary function tests
PCEF = peak cough expiratory flow
NIV = noninvasive ventilation
SNP = sniff nasal pressure
MIP = maximal inspiratory pressure
FVC = forced vital capacity (supine or erect)
Abnl.nocturnal oximetry = pO_2 < 4% from baseline

Text in bold = evidence-based
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