



# THE CARE OF THE PATIENT WITH AMYOTROPHIC LATERAL SCLEROSIS: MULTIDISCIPLINARY CARE, SYMPTOM MANAGEMENT, AND COGNITIVE/BEHAVIORAL IMPAIRMENT

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 multidisciplinary care, symptom management, and the treatment of cognitive/behavioral impairment.

*Please refer to the full guideline at [www.aan.com](http://www.aan.com) for more information, including the AAN's definitions of the levels of recommendations and classifications of evidence.*

## BREAKING THE NEWS

### How should a physician tell patients that they have ALS?

<b>Insufficient evidence</b>	There is insufficient evidence to support or refute any specific method of disclosing the diagnosis in ALS ( <b>Level U</b> ).
<b>Clinical context</b>	Useful strategies have been developed for disclosing a diagnosis of cancer (see appendix e-1 of the published guideline).

## MULTIDISCIPLINARY CARE

### Does multidisciplinary management improve outcomes?

<b>Good evidence</b>	Specialized multidisciplinary clinic referral should be considered for patients with ALS to optimize health care delivery ( <b>Level B</b> ) and prolong survival ( <b>Level B</b> ).
<b>Weak evidence</b>	Specialized multidisciplinary clinic referral may be considered to enhance quality of life (QOL) ( <b>Level C</b> ).

## SYMPTOMATIC MANAGEMENT

### What are the most effective treatments for sialorrhea?

<b>Good evidence</b>	In patients with ALS who have medically refractory sialorrhea, botulinum toxin type B (BTxB) should be considered ( <b>Level B</b> ).
<b>Weak evidence</b>	In patients with ALS who have medically refractory sialorrhea, low-dose radiation therapy to the salivary glands may be considered ( <b>Level C</b> ).
<b>Clinical context</b>	In ALS and other diseases, anticholinergic medications are generally tried first to reduce sialorrhea, although effectiveness is unproven. Botulinum toxin has been effective in controlled trials in parkinsonism as well as ALS.

### What pharmacologic measures reduce pseudobulbar affect?

<b>Good evidence</b>	If approved by the US Food and Drug Administration (FDA), and if side effects are acceptable, dextromethorphan/quinidine (DM)/(Q) should be considered for symptoms of pseudobulbar affect in patients with ALS ( <b>Level B</b> ).
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### What pharmacologic interventions reduce fatigue?

<b>Weak evidence</b>	In patients developing fatigue while taking riluzole, once risks of fatigue versus modest survival benefits have been discussed, withholding the drug may be considered ( <b>Level C</b> ).
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### What interventions reduce cramps?

<b>Insufficient evidence</b>	There are insufficient data to support or refute any specific intervention for the treatment of cramps in ALS ( <b>Level U</b> ).
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### What interventions reduce spasticity?

<b>Insufficient evidence</b>	There are insufficient data to support or refute exercise or medication for treating spasticity in ALS ( <b>Level U</b> ).
<b>Clinical context</b>	In multiple sclerosis and cerebral palsy, benzodiazepam, baclofen, dantrolene, and tizanidine are effective in reducing spasticity-related symptoms.

### What pharmacologic interventions reduce depression?

<b>Insufficient evidence</b>	There are insufficient data to support or refute specific treatments for depression in ALS ( <b>Level U</b> ).
<b>Clinical context</b>	There is consensus among experts that depression should be treated in patients with ALS; however, there are no controlled studies of benefit or harm.

### What pharmacologic interventions reduce anxiety?

<b>Insufficient evidence</b>	There are insufficient data to support or refute specific treatment for anxiety in ALS ( <b>Level U</b> ).
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### What pharmacologic interventions reduce insomnia?

<b>Insufficient evidence</b>	There are insufficient data to support or refute specific treatment for insomnia in ALS ( <b>Level U</b> ).
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## COGNITIVE AND BEHAVIORAL IMPAIRMENT

**What is the prevalence and natural history of cognitive and behavioral impairment in ALS?**

**Good evidence** Screening for cognitive and behavioral impairment should be considered in patients with ALS (**Level B**).

**How is cognitive or behavioral impairment in ALS diagnosed?**

**Weak evidence** Screening tests of executive function may be considered to detect cognitive impairment in patients with ALS prior to confirmation with formal neuropsychological evaluation (**Level C**).

**What is the effect of cognitive or behavioral impairment on management of patients with ALS?**

**Insufficient evidence** There are insufficient data to support or refute the impact of cognitive and behavioral impairment on management in ALS (**Level U**).

**What treatments are effective for cognitive or behavioral impairment in ALS?**

**Insufficient evidence** There are insufficient data to support or refute treatment of cognitive or behavioral impairment in ALS (**Level U**).

## COMMUNICATION

**What treatments for dysarthria optimize communication in ALS?**

**Insufficient evidence** There are insufficient data to support or refute treatment to optimize communication in ALS (**Level U**).

## PALLIATIVE CARE

**What treatments reduce pain and dyspnea in the terminal phase of ALS?**

**Insufficient evidence** There are insufficient data to support or refute specific treatments for pain and dyspnea in late-stage ALS (**Level U**).

**Do hospice care, spiritual interventions, or advance directives improve QOL in the terminal phase of ALS?**

**Insufficient evidence** There are insufficient data to support or refute hospice, spiritual care, or advance directives in ALS (**Level U**).

**What is the optimal method of withdrawing both noninvasive and invasive ventilation in ALS?**

**Insufficient evidence** There are insufficient data to support or refute specific strategies for withdrawal of ventilation in ALS (**Level U**).

**Clinical context**

Protocols based on consensus for withdrawal of mechanical ventilation in intensive care units (Class IV) include counseling and symptom control with opioids, benzodiazepines, and anticholinergic medications. We could find no controlled studies in any disease.

This is an educational service of the American Academy of Neurology. It is designed to provide members with evidence-based guideline recommendations to assist the 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, and are 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.

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