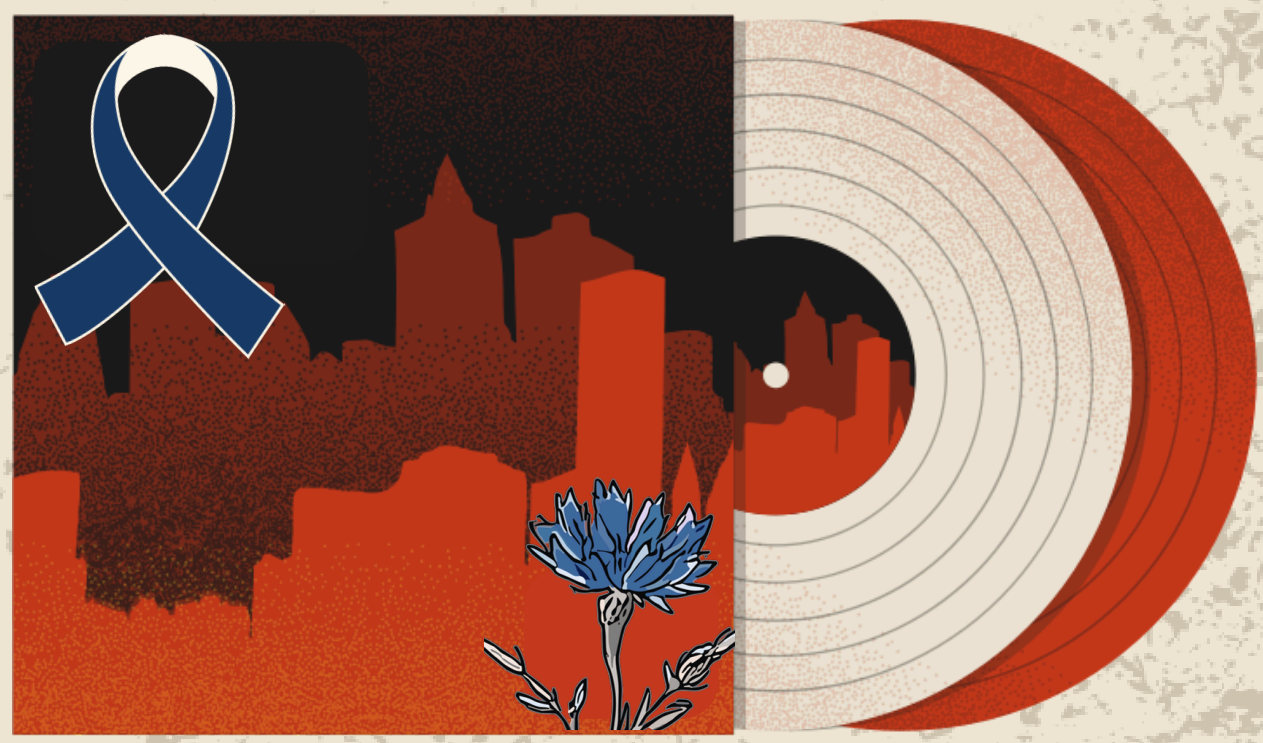
AN SLP'S GUIDE TO

ALS CLINIC

Grace Bailey



2026 | VA LEND LEADERSHIP PROJECT

SPECIAL THANKS TO
DR. KIERA BERRGREN, CCC-SLP
ERIC HANDLER, CCC-SLP
MULTIDISCIPLINARY TEAM AT VCU HEALTH, SHORT PUMP PAVILION

UNTIL THERE IS A CURE, THERE IS CARE.

TABLE OF CONTENTS

ALS

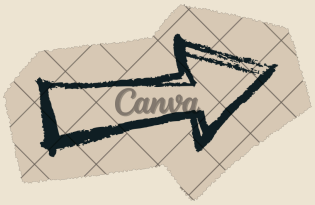
- Progressive neurodegenerative disease affecting nerve cells in the brain and spinal cord.
- Name breakdown: “amyotrophic” means “no muscle nourishment” (leading to muscle wasting), “lateral” refers to areas of the spinal cord affected, and “sclerosis” refers to scarring/hardening of those regions.
- As motor neurons degenerate and die, the brain loses the ability to initiate and control voluntary muscle movement.
- Voluntary muscles affected include those for speaking, swallowing, moving, and breathing.

Prognosis

20% OF PEOPLE WITH ALS LIVE FIVE YEARS, 10% SURVIVE 10 YEARS AND 5% LIVE 20 YEARS OR LONGER

Onset

IN A 2021 MULTICENTER STUDY OF 1,605 ALS PATIENTS, ONSET DISTRIBUTION WAS: 67.0% LIMB, 28.8% BULBAR, AND 4.2% RESPIRATORY



ALS can often mimic other diseases, so the road to a diagnosis may be fraught with various other diagnoses and doctor’s appointments that can lead to some understandable frustration.

The criteria to receive a diagnosis is complex and multifaceted, but signs usually include the following:

- Both UMN + LMN involvement
- Symptoms that spread/progress
- EMG showing denervation in several regions
- No other disease that explains it

PLS

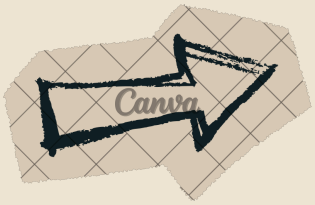
- PLS is a rare, degenerative neurological disorder primarily affecting the upper motor neurons.
- Caused by degeneration of upper motor neurons in the brain and spinal cord, leading to progressive spasticity and weakness.
- Often described as a benign variant of ALS because it progresses more slowly.
- Major difference from ALS:
  - PLS affects only upper motor neurons.
  - Lower motor neurons remain intact, so there is no muscle wasting (amyotrophy).
- Symptoms usually start in the legs, but onset may occur in the arms or bulbar muscles (speech/swallowing).

Prognosis

SLOW, DECADES-LONG PROGRESSION WITH NORMAL OR NEAR-NORMAL LIFE EXPECTANCY

Onset

IN PLS, SYMPTOMS COMMONLY BEGIN IN THE LEGS, BUT ONSET CAN ALSO OCCUR IN THE ARMS OR BULBAR MUSCLES



ALS clinics often also treat people with PLS because the conditions share similar motor symptoms, require overlapping specialists, and benefit from the same multidisciplinary expertise in mobility, spasticity management, speech, swallowing, and long-term care planning.

The criteria to receive a diagnosis is individualistic, but signs usually include the following

- Progressive symptoms of upper motor neuron dysfunction for at least 3-4 years
- No lower motor neuron involvement on exam or EMG
- Symptoms that begin in legs, arms, or bulbar muscles and gradually spread
- EMG remains normal or near-normal, with no signs of active denervation

Pseudobulbar Affect

Pseudobulbar affect is a neurological condition that can sometimes occur in those with ALS, particularly those with the bulbar onset variation. The condition manifests as uncontrollable crying and laughing, often with no apparent reason for doing so. It can also manifest as laughing and crying at the same time.

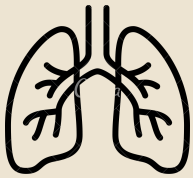
"The crying episodes in PBA are brief and not necessarily related to sadness. Other symptoms of depression, such as appetite loss or sleep disturbances, are usually absent in people with ALS who have PBA" (Strachan, 2023)

"Patients with ALS showed a PBA prevalence of 38.5%" (Nabizadeh et al., 2022).

The Multidisplinary Team

"A multidisciplinary team comprises professionals from different disciplines who come together to provide comprehensive care and support for patients or clients. The team works collaboratively, each member bringing their specialized skills and expertise to develop and implement coordinated services and care plans tailored to the individual's unique needs" (Meridian Team, 2024).

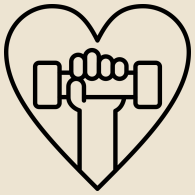
WHO'S ON THE TEAM?



RESPIRATORY THERAPIST

NEUROLOGIST

OCCUPATIONAL THERAPIST



PHYSICAL THERAPIST

GENETIC COUNSELOR

DIETICIAN

ALS SOCIETY REPRESENTATIVE

SOCIAL WORKER

NURSE NAVIGATOR

SPEECH-LANGUAGE PATHOLOGIST

Registered Dietician
RD

SCOPE OF PRACTICE

1. Track weight, BMI, and percent weight change; intervene early
2. Prevent/manage weight loss with high-calorie, high-protein strategies
3. Assess oral intake, appetite, meal fatigue, and nutrition-impact symptoms
4. Recommend safer, easier-to-swallow foods and hydration options
5. Provide calorie-dense recipes, supplements, and fortified meals
6. Manage constipation, reflux, bloating, early satiety, and dehydration
7. Initiate and guide PEG/G-tube timing discussions
8. Support caregiver burden with meal-prep shortcuts and resources
9. Maintain quality of life by maximizing enjoyment and safety of eating

WHAT MAY BE NOTICED

1. Unintentional weight loss; clothes fitting looser
2. Difficulty finishing meals; chewing fatigue
3. Taking longer to eat; prolonged mealtimes
4. Avoidance of hard, dry, or mixed textures
5. Signs of dehydration (dark urine, dry mouth, thick mucus)
6. Reduced appetite or early satiety
7. GI issues: constipation, reflux, bloating, nausea
8. Difficulty maintaining upright posture when and after eating
9. Avoidance, fear, or denial around PEG/G-tube discussion
10. Caregiver stress or overwhelm related to meals

TYPICAL RECOMMENDATIONS

1. **Increase calories with add-ins (butter, oils, nut butters, cream, cheese).**
2. **Eat small, frequent meals to reduce fatigue.**
3. **Add high-calorie, high-protein shakes or supplements between meals.**
4. **Prioritize hydration with water, broths, electrolyte drinks, or gels.**
5. **Use meal-prep shortcuts: pre-chopped foods, simple menus, ready-to-eat options.**
6. **Schedule meals around energy peaks and medications.**
7. **Manage constipation with fiber, fluids, and daily routine.**
8. **Consider timing of PEG/G-tube placement when weight loss or intake decline begins**

one smart cookie
Special thanks to Rebecca Rhodes, RD

RD & SLP

The RD and SLP work closely together in multidisciplinary clinics, sometimes even going into patient's rooms together. Swallowing safety and nutritional status are intertwined, and the RD and SLP's skills complement each other during a patient's care. The SLP assesses swallowing function, identifies aspiration risks, recommends safe textures and liquid consistencies, and monitors how fatigue and respiratory changes affect eating. The RD then uses this information to design nutrition plans that maintain weight, hydration, and quality of life while aligning with the patient's swallow abilities. Together, they coordinate strategies such as meal pacing, texture modifications, supplement use, and timing of PEG placement. They also collaborate during meal observations, check-in frequently as symptoms progress, and provide unified education to patients and caregivers. By combining safety-focused swallow management with nutrition-focused intervention, the SLP and RD ensure that patients receive cohesive, comprehensive support throughout the course of ALS.

WORKING TOGETHER

Tips for Working in Tandem:

- Ask each other questions when boundaries feel unclear instead of assuming
- Be clear about roles from the beginning. If you're new to the environment, confirm roles at the beginning to avoid conflict later.
- Communicate after every swallow eval or significant nutrition change
- Clarify who leads conversations about PEG and when each clinician joins in
- Present a united front during caregiver education to reduce overwhelm and confusion
- Notify each other promptly when new red flags appear (weight loss, choking, prolonged meals)

JOINT COUNSELING

Topics both clinicians reinforce:

- **S**afe independence
- Fatigue management
- Maintaining food joy
- Caregiver support
- Preparing for changes without removing autonomy

Messages that both reinforce:

- Eating should be safe and efficient
- Fatigue affects swallow safety
- Weight loss is not "normal aging"
- Pleasure and autonomy in eating still matter
- PEG does not eliminate oral intake for pleasure

Respiratory Therapist

A respiratory therapist (RT) plays a crucial role in ALS care by monitoring breathing, supporting airway clearance, and helping patients maintain comfort and safety as respiratory muscles weaken. The RT performs pulmonary function testing–especially measures like vital capacity (VC), forced vital capacity (FVC), and slow vital capacity (SVC)–to track diaphragm and chest wall function over time. **These numbers guide care across the team; many clinics begin discussing non-invasive ventilation when VC or FVC approaches 50% of predicted, and values nearing 30% indicate significant respiratory compromise requiring close management.** RTs also assess MIP and MEP, which measure inspiratory and expiratory muscle strength and help determine cough effectiveness and overall respiratory muscle weakness in ALS. In addition, the RT educates patients and caregivers, introduces and adjusts non-invasive ventilation (BiPAP), teaches breath-stacking and cough-assist techniques, and helps manage secretions and nighttime breathing symptoms. They work closely with neurology, the SLP, and the dietitian, since changes in respiratory strength affect swallowing safety, mealtime endurance, hydration, and overall quality of life.

MECHANICAL OPTIONS

BIPAP

PORTABLE VENTILATOR

COUGH ASSIST MACHINE

SUCTION MACHINE

If some patient are able to get off their ventilator for only small periods of time, advise smaller, more frequent, and calorie-dense meals.

Some patient may not be able to be off their machine for very long or at all to speak more clearly. A voice amplifier may be useful; however, it will amplify all the noises (machine noise, burping, coughing, etc.). Some amplifiers have addressed this.

Social Worker

- Emotional support for patients and caregivers
- Help with insurance, disability, and benefits
- Connect families to resources, grants, and support groups
- Assist with advance care planning and end-of-life decisions
- Coordinate home care, equipment, and community services
- Address caregiver stress and provide coping strategies
- Advocate for patient needs across the care team

Clinical Psychologist

- Support diagnosis acceptance and adjustment.
- Guide PEG and other major care decisions.
- Assess and manage depression, anxiety, and caregiver strain.
- Help clarify goals of care and quality-of-life priorities.
- Facilitate family communication and advance planning.
- Monitor cognitive/behavioral changes affecting safety or judgment.
- Provide caregiver coping strategies and burnout support.

Genetic Counselor

- Helps you understand family history, ALS genetics, and what testing can show.
- Reviews the benefits, risks, and limits of genetic testing before you decide.
- Explains what positive, negative, or uncertain results mean.
- Supports you through the emotional, social, and family implications of testing.
- Helps arrange testing, consent, and insurance/cost questions if you choose to proceed.
- Interprets results and discusses what they mean for you and your relatives.
- Provides guidance on family planning, talking to relatives, and next steps.

FYI

C9orf72 is the most common genetic cause of ALS and frontotemporal dementia (FTD). It's inherited in an autosomal dominant pattern, meaning first-degree relatives have a 50% chance of carrying the expansion, though not everyone who carries it will develop symptoms. The gene change can lead to ALS, FTD, or a combination of both, and is sometimes associated with behavioral or personality changes when FTD features are present. A positive C9 result has important implications for family members but cannot predict when symptoms will start, what symptoms will appear, or how quickly disease may progress, which is why genetic counseling is essential before and after testing.

Nurse Navigator

A nurse navigator is often the central point of coordination in ALS care, helping patients and families move smoothly through a complex medical system. They track symptoms between clinic visits, manage communication among team members, and ensure that concerns reach the right provider quickly. The nurse navigator assists with medication questions, equipment orders, insurance issues, referrals, and understanding test results, while also monitoring for urgent changes in breathing, swallowing, mobility, or overall health. They provide patient and caregiver education, reinforce safety strategies recommended by the team, and support advance care planning as needs evolve. Because they maintain consistent, ongoing contact with the family, the nurse navigator plays an essential role in continuity of care, emotional support, and helping patients feel guided rather than overwhelmed throughout the course of ALS.

Physical Therapist

A physical therapist in an ALS clinic focuses on maintaining mobility, comfort, and safety as the disease progresses. They assess strength, balance, gait, and fall risk, then recommend exercises that are gentle and energy-conserving. They fit and adjust mobility aids, braces, and orthotics, help manage spasticity and pain, and teach safe transfer techniques. Their role centers on keeping patients moving as safely as possible for as long as possible, while preparing them and their caregivers for upcoming physical changes.

Occupational Therapist

An occupational therapist in an ALS clinic helps patients stay as independent, safe, and efficient as possible in daily life. They assess how weakness, fatigue, or fine-motor changes affect tasks like dressing, writing, cooking, and bathing, then recommend adaptive strategies or equipment to make those tasks doable. They address energy conservation, home modifications, and positioning, and they prepare patients and caregivers for future needs by introducing tools like built-up utensils, bathroom safety equipment, and hands-free technology. Their goal is to keep meaningful activities accessible for as long as possible.

ALS FOUNDATION REP

Results of an Interview with Meg Greene, ALS clinic liaison

Role Summary

- Serves as the main point of contact between patients/families and ALS Association resources.
- Assesses patient needs during clinic visits and connects them with appropriate supports.
- Focuses on meeting patients where they are and avoiding information overload.

Equipment & Financial Resources

- Coordinates access to:
- DME Loan Closet - equipment not covered by insurance.
- AAC Loan Closet - communication devices.
- Loaner ramps and other safety equipment.

> *"Don't overwhelm them by going into too much detail. Meet them where they're at. Listen to what they're dealing with."*

Registration & Documentation

- Patients must be registered with the ALS Association to access supports.
- Liaison assists with:
  - Account creation within the internal documentation system.
  - Navigating the order portal for equipment and services.

Grants

- Quality of Life Grant
  - $1,500 annually (government-funded; changing to half next year).
  - Can be used for any need.
  - No receipts or follow-up required.
- Hoffman Travel Grant
  - $1,000 annually.
  - Supported by the Hoffman family's historic $58M donation.
  - Assists with clinic travel and access.

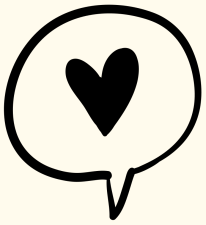
ALS Registration Packet

Association Resources and Support

Support Groups

- All groups offered virtually.
- Includes: gender-specific groups, caregiver groups, national Spanish-language group, additional regional groups (e.g., Hampton Roads partnership).

"I feel very fortunate to be here. When I talk to the people here, you guys, the families, the setup, the flow, even though it may not seem completely structured, I rely on the team."



Your Turn

Think comfort and safety care - not rehab.
Less exercises, more adaptation.
Quality of life is the driving force.

Sample Questions List

1. How do you prefer to be called?
2. How has eating been going for you?
3. How long do meals take?
4. Any foods you've been avoiding?
5. How has hydration been?
6. How are pills going for you?
7. Has your weight stayed stable?
8. Are we brushing our teeth? Are you a flosser?

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1. Have you noticed any changes in your speech or voice?
2. Any communication holes we need to fill?
3. Do you have a smartphone? What do you use it for?
4. Have we discussed voice/message banking?
5. How do you get help if you need it?

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1. Have you noticed any changes in your memory, your thinking skills, attention/focus?

Questions List Reasoning

- **How do you prefer to be called?**
- **How has eating/swallowing been going for you?**
  - Sometimes the pt may answer that everything's fine, but the following questions may jog their memory or highlight an issue they haven't noticed yet.
  - You may need to clarify by asking if they've been choking or coughing recently.
- **How long do meals take?**
  - This question helps highlight a fatigue problem or potential dysphagia which may affect calorie intake, socializing, and frustration levels.
- **Any foods you've been avoiding?**
  - This question highlights particular textures that may be causing problems. For example, a pt may be avoiding bread because they choke on it, which could indicate an emerging dysphagia. You could educate them on the food continuum and encourage them to add softeners like butter, gravy, oil, et cetera.
- **How has hydration been?**
  - In ALS, staying hydrated can get harder without people realizing it. Changes in swallowing, fatigue, or hand weakness can make drinking enough a challenge. When hydration drops, secretions get thicker, breathing feels harder, and swallowing safety can actually get worse. So checking in on fluid intake helps you catch early signs of trouble and make sure we're supporting them before it snowballs.

Questions List Reasoning

- **How are pills going for you?**
  - Sometimes, a patient won't complain of an eating/swallowing problem, but they will endorse that pills can be difficult. Educate them on the food continuum as they may benefit from swallowing their pills with a puree (triggers a stronger swallow) and/or asking the pharmacist for a different method of delivery (many big vitamins come in a liquid form). Remind them that they could crush the pills into a puree as well, but they will need to check with the doctor and the pharmacist, in case there is a contraindication.
- **Has your weight stayed stable?**
  - People often lose weight without trying due to increased energy needs or subtle swallowing changes. That weight loss can actually speed up weakness and make breathing and swallowing harder. So checking in on whether their weight has been steady helps you catch red flags early and adjust nutrition or swallowing supports before it becomes a bigger issue.
- **Are we brushing our teeth? Are you a flosser?**
  - **O**ral care becomes more challenging as hand strength, coordination, or fatigue change in ALS. If brushing gets harder, bacteria and residue can build up, which makes swallowing less safe and increases the risk of respiratory infections. Checking in about oral care helps you see if they need adaptive tools, caregiver support, or modified routines so they can keep their mouth healthy and swallowing safer.

Swallow

- All swallow muscles **gradually weaken**, starting subtly and progressing continuously.
- Tongue strength typically reduces first, making chewing and bolus control slower and less efficient.
- Lip, jaw, and cheek muscles weaken, causing spillage, pocketing, and early meal fatigue.
- Pharyngeal muscles weaken, slowing the swallow trigger and reducing airway protection and clearance.
- Laryngeal elevation and closure weaken, increasing risk of penetration and silent aspiration.
- Respiratory muscles weaken, making the cough too weak to clear aspiration.
- Fatigue worsens everything, so performance drops within minutes of starting a meal.

Clinical Indicators

- Coughing with thin liquids, especially water.
- Taking longer to chew, especially tougher foods.
- Food sitting in the cheek or needing reminders to clear it.
- Chewing looks "tired" – slow, effortful, or incomplete.
- Needing multiple swallows to get one bite or sip down.
- Wet or gurgly voice after eating or drinking.
- Increased throat clearing during meals.
- Avoiding mixed textures (cereal + milk, salads).
- Meals taking much longer – sometimes double or triple the time.
- Fatigue halfway through a meal, needing breaks or stopping early.
- Unexplained weight loss despite eating "the same amount."
- Choking episodes or looking fearful/hesitant around food.
- Not talking while eating anymore because it feels harder to coordinate.
- Preferring softer foods without being aware they're compensating.
- Getting short of breath during meals or needing to pause to breathe.
- Eating less overall, saying they're "not hungry" (actually too fatigued).

Reflux

Ask if they're coughing in between meals or at night. These symptoms are likely due to reflux, which may need to be medically or behaviorally managed with reflux precautions.

The Modified Barium Swallow Test

- Some ALS patients do get a modified, but we don't order them routinely in our particular clinic.
- ALS swallow decline is typically predictable, so the study often doesn't add new or meaningful information.
- Our recommendations (texture changes, pacing, airway protection, PEG timing) stay the same with or without imaging.
- Getting to the radiology facility can be exhausting, difficult, and sometimes unsafe for patients with mobility or respiratory issues.
- If a modified won't change the plan, it isn't worth the travel, transfer, or fatigue of the appointment.
- We do order one if something seems unusual, sudden, or inconsistent with typical ALS swallowing patterns.
- Otherwise, ongoing clinical monitoring in the clinic is the most practical, compassionate approach.

FOOD/DRINK CONTINUUM

Patients often benefit from an explanation of the "food/drink continuum."

On either side of the continuum, we have things that are typically more difficult to swallow. On one side are things like steak, chicken, and salad; on the other, thin liquids like water or tea. As we move closer to the center of the continuum, foods start to looks a little more like puree: things like applesauce, pudding, or yogurt. These foods are typically easier to swallow, so if these things on either side of the continuum are giving you trouble, try making your food closer to a puree. It doesn't have to be applesauce, but think "moist" or "soft" which will make your bite a little more cohesive. For example, add more gravy to the chicken, extra butter to potatos, et cetera.

Patient often benefit from the food continuum concept when you recommend taking pills with a puree, instead of water, if pills are getting "stuck" or the pills cause a choking event.

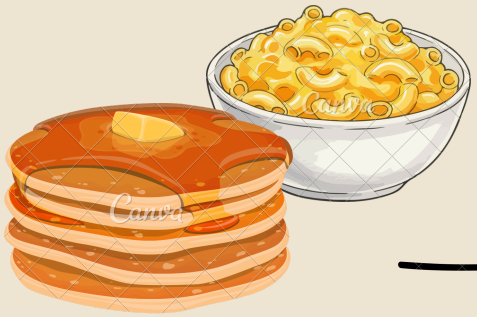
Foods closer to the center of the continuum are often easier to swallow and often trigger a harder, more effective swallow, carrying the pill with less incidence. It doesn't have to applesauce or yogurt, but maybe just something thicker, like milk or a smoothie.

Forough, A. S., Lau, E. T. L., Steadman, K. J., Cichero, J. A. Y., Kyle, G. J., SerranoÂ Santos, J. M., & Nissen, L. M. (2018). A spoonful of sugar helps the medicine go down? A review of strategies for making pills easier to swallow. Patient Preference and Adherence, 12, 1337-1346. https://doi.org/10.2147/PPA.S164406

Steele, C. M., Alsanei, W. A., Ayanikalath, S., Barbon, C. E. A., Chen, J., Cichero, J. A. Y., ... Wang, H. (2015). The influence of food texture and liquid consistency modification on swallowing physiology and function: A systematic review. Dysphagia, 30(2), 2-26. https://doi.org/10.1007/s00455-014-9578-x

FOOD/DRINK CONTINUUM

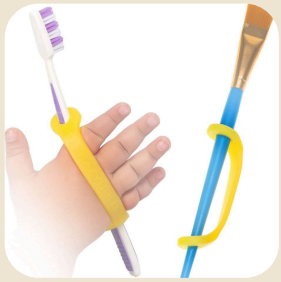
Tough Solids

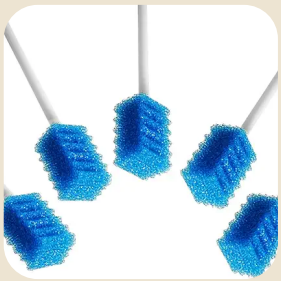


Puree

Thin Liquids

Adaptive Menu





Speech

In distal-onset ALS, speech is usually normal at the beginning because weakness starts in the limbs, not the bulbar muscles. As the disease progresses and bulbar involvement eventually appears, people typically develop:

- Quieter voice (reduced loudness)
- Mild breathiness or a slightly weak-sounding voice
- Subtle articulation imprecision (especially lingual sounds)
- Shorter phrases as respiratory support decreases
- Occasional strain or effortfulness when UMN signs emerge
- Gradual changes–nothing abrupt early on

Spastic Dysarthria Examples

Mixed Flaccid-Spastic Example

Sometimes caregivers become the primary communicator, even when the patient is still able to express themselves. Ensure that you continuously include the patient for their endorsement.

In bulbar-onset ALS, speech changes are one of the very first symptoms. People typically present with:

- Slurred articulation (especially tongue-heavy sounds)
- Breathy or rough voice
- Reduced loudness or hypophonia
- Early hypernasality from velopharyngeal weakness
- Rapid fatigue during speaking
- Faster progression to reduced intelligibility

Communication

How do you get help when you need it?

Sometimes they haven't thought of this before.

Ask them where their phone is (not on your person?)

There are options!

Sometimes they are attended 24/7, so it may not be necessary.

Wireless Doorbell

Texting 911 is sometimes an option, depending on the county

Home systems that can be orally activated

LifeAlert System

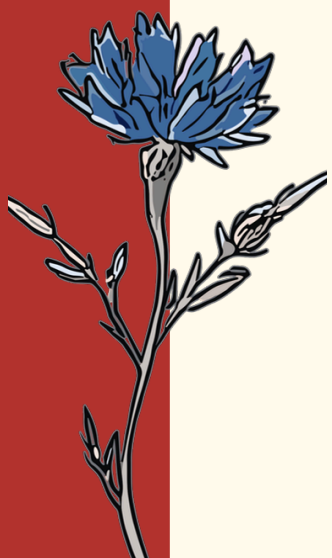
Gizmo Pal

Chin Switch (and various others, via OT)

Communication

1. Let the patient know that there are many options out there to support communication. Avoid a full discussion at the time of diagnosis unless time is of the essence or they're very interested
2. An AAC eval should be able to fit them with the best device for their needs and skills.
3. Mention that being fitted for a device is better to do sooner rather than later to learn (once they've made the decision) and be comfortable with the device
4. Send them information to review and continue to follow-up, if they're interested

Speech Assistant app is a great example to have handy to show them a little of what is possible. It's fully customizable with a choice of automated voices, font sizes, button sizes, and more. There's a lite version for Android, although Android does not allow you to do phone calls. iPhone can use the app when speaking on the phone, which can be very appealing to patients, although there's an outright, one-time fee for the app..



Speech Assistant App

Voice/Message Banking

What is it?

- Voice banking: Recording a large set of phrases to create a synthesized voice that sounds like the person.
- Message banking: Recording specific, meaningful phrases in the person's own natural voice (e.g., "I love you," jokes, nicknames) that can be played back exactly as recorded.
- Double-banking: Doing both–so you get a synthesized voice and personal messages (Boston Children's Hospital Augmentative Communication Program, n.d.)

Why It Matters

- Helps preserve personal identity and emotional connection when speech declines.
- Gives the person control and autonomy over how they sound later.
- Reduces distress for families when natural speech is lost.
- Ensures AAC devices can reflect personality, tone, and cultural/linguistic style.

Potential Barriers

- Fatigue–recording can be tiring
- Emotional weight of planning for future loss
- Technology intimidation
- Time and cost (some programs require fees).).

Patient Education

- You'll likely need to send information virtually. Continuously follow-up with it if they are willing.
- Encourage them to consider it as a proactive step, even if their voice is not currently different. They may thank themselves later.
- Don't assume that they aren't tech-savvy enough to do anything with the voice banking.
- Asking about what type of phone they have and what they use it for will help figure out a ballpark for their technology limits.

Options

Acapela

Speak Unique

The Voice Keeper

Model Talker

Team Gleason

Veteran Services

Apple Personal Voice

My Message Banking

Voice Cloning

Voice Cloning Assistance

Compare/contrast Chart

Conan O'Brian Podcast

Cognition

- Not everyone who is diagnosed with ALS will experience cognitive and/or behavioral changes.
- Cognitive or behavioral impairment occurs in up to 50-65% of individuals with ALS. About 15% meet criteria for full frontotemporal dementia.
- If these changes do occur, they often present as FTD (frontotemporal dementia) and may be diagnosed with ASD-FTD (Amyotrophic Lateral Sclerosis-Frontotemporal Spectrum Disorder).
- Common genetic component: C9ORF72
- Cognitive/behavioral changes may occur before or after motor changes.
- "ALS-FTSD patients have a worse prognosis and shorter survival rates than patients with ALS"
- FTD is different than Alzheimer's. Their criteria are distinct.
- Bulbar onset (speech/swallow involvement) correlates with increased risk of cognitive/behavioral changes
- Clinical Impact: reduced survival rate, potentially poor compliance, patient/caregiver frustration and fear, poor safety awareness, inability to make important decisions, and reduced driving skills.
- Executive dysfunction (planning, organization, inhibition, cognitive flexibility) is the most common cognitive domain affected.
- Behavioral symptoms include apathy, loss of empathy, irritability, disinhibition, and reduced insight.
- Language impairments may occur, often mirroring the non-fluent/agrammatic variant of primary progressive aphasia (PPA).
- Social cognition deficits (emotion recognition, theory of mind) can appear even when traditional cognitive testing is normal.

The ALS-CBS Screener

Cognitive Behavioral Screener

The ALS Cognitive Behavioral Screen (ALS-CBS) is a quick, clinic-friendly tool used to detect cognitive and behavioral changes in people with ALS, especially those on the ALS–FTD spectrum. It has two parts: a cognitive section that assesses attention, executive function, fluency, and working memory through brief tasks, and a behavioral section completed by a caregiver or clinician to identify changes in personality, social behavior, judgment, or apathy. It's designed to flag whether someone may need a more comprehensive neuropsychological evaluation, and it helps the team understand how cognitive or behavioral symptoms might impact safety, communication, and decision-making.

Note that giving the ALS-CBS screener at a patient's first visit may not be reflective of their true baseline as they may have just received their diagnosis, life-changing news that can result in a wide variety of emotions that will skew results and cause more harm than good.

ALS CBS
ALS Cognitive Behavioral Screen

Susan C. Woolley, Ph.D.

Patient Id: ____________ **DOB/Age:** ________ **Gender:** ____
Onset Date: __________ **FVC:** ________ **Education:** ____
Onset Region: bulbar, arm, leg, trunk, respiratory (circle one) ☐ Mark if pt response were written, attach sheet

---

HAND PAGE 2 TO CAREGIVER

Attention

a. Commands: *I am going to say some commands. Please listen carefully and then do what I say. (If patient is unable to indicate with finger, movement can be substituted with eyes, arm or other means)*
1. Point/indicate (with your finger) to the ceiling and then to your left.
2. Touch your shoulder, point to the floor, and then make a fist.

errors: 0 | 1+
Score (circle): 1 | 0

b. Mental Addition/Language: *I am going to say some phrases. I want you to tell me the number of syllables in each phrase. For example, "the table" has 3 syllables. (Repetition of each phrase is allowed once)*
1. The weather is nice. (correct response: 5) answer ____
2. Tomorrow will be sunny. (correct response: 7) answer ____

errors: 0 | 1+
Score (circle): 1 | 0
(score 0 if >20 seconds on either)

c. Eye Movements: *Saccades and Antisaccades*
# of Correct Saccades out of 8: ____/8 Score: 8/8 = 1 points, ≤7/8 = 0 points
# of Correct Antisaccades out of 8: ____/8 Score: 8/8 = 2 points, 7/8 = 1 points, ≤6/8 = 0 points

/5

Concentration

I am going to say some numbers. After I say them, I want you to say them to me backwards, or in reverse order. For example, if I say 3-6, you would say 6-3. (If written, do not allow pt to write forward span. Discontinue after failure on two consecutive trials)

		Correct	Incorrect			Correct	Incorrect
2-9	(9-2)	__	__	7-5-6-4	(4-6-5-7)	__	__
6-8	(8-6)	__	__	5-8-1-9	(9-1-8-5)	__	__
3-7-2	(2-7-3)	__	__	8-2-5-9-3	(3-9-5-2-8)	__	__
5-8-1	(1-8-5)	__	__	5-7-6-3-9	(9-3-6-7-5)	__	__

Maximum Span Correct: (Enter score) /5

Tracking/Monitoring

a. Months: *Please say the months of the year backwards, starting with December. (circle omissions/mark repetitions & intrusions)*

Dec Nov Oct Sep Aug Jul Jun May Apr Mar Feb Jan

errors: 0 | 1 | 2+
Score (circle): 2 | 1 | 0

b. Alphabet: *Please say/write the alphabet for me. (mark uncorrected errors, omissions or intrusions)*

A B C D E F G H I J K L M N O P Q R S T U V W X Y Z

errors: 0 | 1+
Score (circle): 1 | 0

c. Alternation Task: *I want you to alternate between numbers and letters, starting with 1-A, and then 2-B, 3-C, and so on. Please continue from there, alternating between number-letter, number-letter, in order, without skipping any until I tell you to stop. (Errors: Any mistake in sequencing, i.e., 7-H or 8-I)*

4-D 5-E 6-F 7-G 8-H 9-I 10-J 11-K 12-L 13-M

errors: 0 | 1 | 2
Score (circle): 2 | 1 | 0

/5

Initiation and Retrieval *Say (write) as many words as you can starting with the letter F, as quickly as you can, in 1 minute. (Show pt Fluency Rules) You cannot say/write the names of people, places or numbers. Please do not say/write the same word with just a different ending, like truck, trucks. (S words can be substituted for F words). Errors: repetitions, rule violations*

1.	________	9.	________	17.	________
2.	________	10.	________	18.	________
3.	________	11.	________	19.	________
4.	________	12.	________	20.	________
5.	________	13.	________		
6.	________	14.	________		
7.	________	15.	________		
8.	________	16.	________		

correct words: >12 | 12-9 | <9 | ≤4
Score (circle): 3 | 2 | 1 | 0*

plus

errors: 0 | 1 | 2+
Score (circle): 2 | 1 | 0

*If ≤4 words, total verbal fluency score = 0 regardless of # of errors

/5

TOTAL SCORE /20

ALS CBS
ALS Cognitive Behavioral Screen

Susan C. Woolley, Ph.D.

ALS Caregiver Behavioral Questionnaire

These questions pertain to possible changes that you have noticed since the onset of ALS symptoms. As best you can, consider changes that are unrelated to physical weakness. For example, question #1 asks about interest in activities. If the person can no longer play tennis but still seems interested in it (i.e. talks about it, watches it on television), then you would circle 3 for no change in level of interest.

If the person has always had the trait in question, please respond No Change, since there has been no change over time.

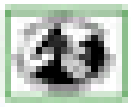
Compared to before ALS, does he/she:

	No Change	Small Change	Medium Change	Large Change
1. Have less interest in topics/events that used to be important to them?	3	2	1	0
2. Show little emotion, or seem less responsive emotionally?	3	2	1	0
3. Seem more agreeable or pleasant than in the past with fewer worries?	3	2	1	0
4. Fail to think things through before acting?	3	2	1	0
5. Seem more withdrawn from others but not sad?	3	2	1	0
6. Get confused or distracted more easily?	3	2	1	0
7. Have less ability to deal with frustration or stress?	3	2	1	0
8. Seem less concerned about the feelings or concerns of others than before?	3	2	1	0
9. Get angry or irritable more easily than before?	3	2	1	0
10. Seem more sarcastic or childlike than before?	3	2	1	0
11. Eat more or have a new preference for particular foods (i.e. sweets)?	3	2	1	0
12. Have more trouble changing opinions or adapting to new situations?	3	2	1	0
13. Show less judgment or more problems making good decisions (i.e. regarding safety, finances, etc)?	3	2	1	0
14. Have less awareness of obvious problems or changes, or deny them?	3	2	1	0
15. Have new problems with language, such as saying the wrong word more often, making up new words, or declines in spelling ability?	3	2	1	0

TOTAL SCORE: _____/45

The following questions relate to current symptoms, not changes over time:

Do you think your loved one:	**YES**	**NO**
• Seems depressed on most days?	[ ]	[ ]
• Seems anxious on most days?	[ ]	[ ]
• Seems extremely fatigued on most days?	[ ]	[ ]
• Suffers from unexpected crying or laughing spells?	[ ]	[ ]



Susan C. Woolley, Ph.D.

Eye Movement Instructions

Saccades: *I am going to hold my fingers up. Please keep your head straight and look at me. When I wiggle a finger, I want you to look at that finger and then look back at me (examiner should execute this eye movement themselves to demonstrate). Look at my finger by moving your eyes only, trying to keep your head still. Each time I wiggle a finger, look at it and then back to me. (Do 2-3 trials with the patient as practice) We will do that a few times. Ready?* (Do 8 random trials, pause for 1-2 seconds between each trial).

Antisaccades: *Good, next I am going to wiggle a finger again, but this time, I want you to look AWAY from the finger that moves. For example, if I move this finger (wiggle one) then I want you to look at the other finger, not the one that moves, ok?* (Examiner should demonstrate for patient) *Let's try it (do 2-3 trials). Just like before, try to keep you head still and just move your eyes. After each one, look back at me. Ready?* (Do 8 random trials, pause for 1-2 seconds between each trial).

FLUENCY RULES

NO NAMES OF PEOPLE

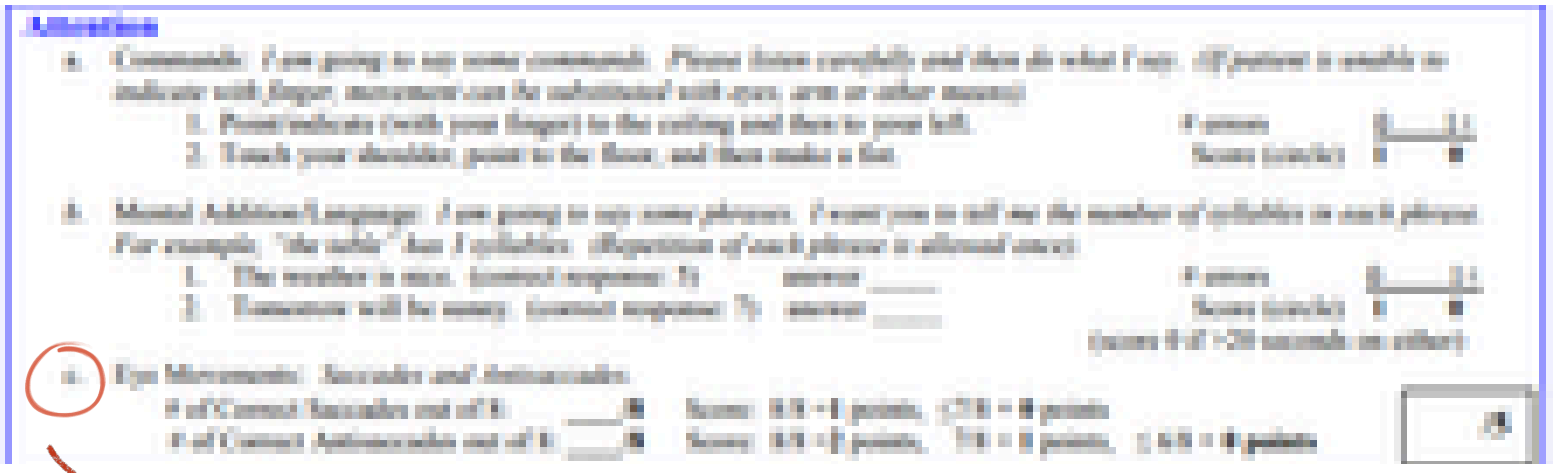
NO NAMES OF PLACES

NO NUMBERS

DO NOT USE SAME WORD WITH DIFFERENT ENDING

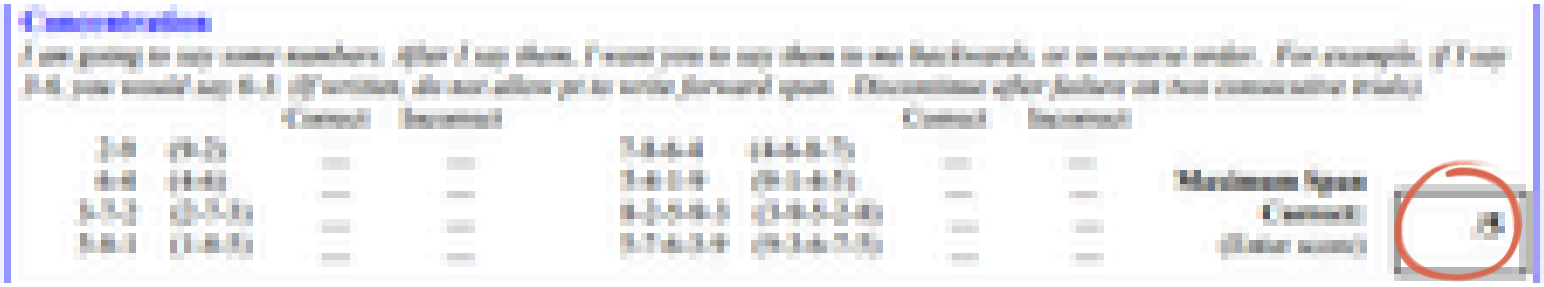
SCREENER WALK-THROUGH

Ask about their highest level of education (doesn't change the score, but informs the severity).

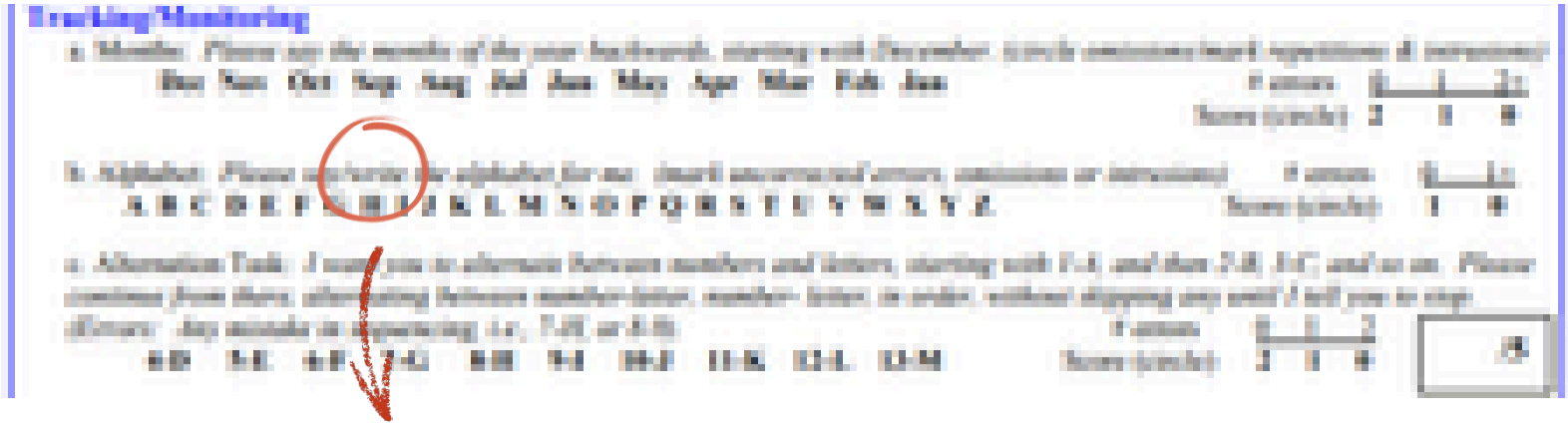


Hold your two index fingers out. Ask if they can see both of them in their field of vision. Have them look at your nose then look to the finger that wiggles, then back to your nose. Anti-saccades are when they're instructed to look at the finger that doesn't wiggle.

Clinical indicator for potential FTD: Pt may stare too long at your finger and take too long to look back at your nose, even with prompting.



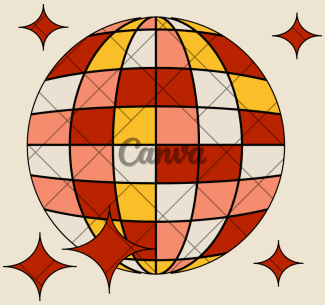
You total this score by counting the maximum number of digits they can list in their highest correct trial. For example, if they successfully say 1-8-5 but stumble on the four digit trial, they would score a 3.



The pt can also write answers, if that is there communication modality.

Be sure to not dismiss proper nouns that can also be common ones (ie., "Frank" versus "[to be] frank").

Sample Note



Dx: ALS (put "pending" if it's not official)
Attended by wife

Speech: Patient presents with mild spastic dysarthria with mild hypophonia marked with intermittent mild imprecisions. Their intelligibility is judged to be >95% in a room with minimal distractions and noise. Continue to write about their abilities.

Language: Expressive/receptive language skills appear to be WFL. These skills were informally assessed. Note that sometimes these skills can be affected by cognition changes or stroke.

Communication: Patient presents with mild spastic dysarthria with mild hypophonia marked with intermittent mild imprecisions. Their intelligibility is judged to be >95% in a room with minimal distractions and noise. Continue to write about their abilities.

Cognition: Cognitions appears to be WFL; these skills were informally assessed. Here is where you can talk about the ALS-CBS screener, note frequency of anomia or other incidents, et cetera.

Swallow: Patient has no concerns at this time. OR mention frequency of choking episodes, how much nutrition is by mouth/by PEG, interest in PEG, education about PEG, where food or liquids or pills are getting stuck, education on puree, texture modifications, urine color, meal frequency and average length, weight stability

Recs: List recommendation is the same order as they appear in your note.

Medicines

You may see these medicines or hear about them in relation to the deficits you treat. Being familiar with medicines will help you identify what is being treated as well as potential side-effects that affect your treatment. The list is for your edification, as you know that medication alterations are not in your scope of practice; however, the neurologist may trial a different drug if the side effects or efficacy prohibit quality of life. Additionally, some drugs can be changed from pill to liquid or crushed, depending on their release pattern; consult or have patient consult pharmacist and neurologist.

ALS DRUGS

Qalsody (Tofersen) - lumbar puncture (only really helpful for SOD1 mutations)
Radicava (Edaravone) - IV or oral suspension (can be administered by mouth or via feeding tube)
Riluzole (Exservan, Rilutek, Tiglutik) - tablet, thickened liquid (Tiglutik), or oral film (Exservan)
Nuedexta (Dextromethorphan HBr and Quinidine Sulfate) - capsule, help treat pseudobulbar affect

THICK SECRETIONS

Guaifenesin (OTC) - Thins mucus; easy to take
Papaya Enzyme (papase) - Helps dissolve thick secretions
Acetylcysteine (nebulized) - For very thick mucus; may irritate airway
Potassium Iodide (SSKI) - Increases thin saliva to reduce thickness; slow onset (up to 2 weeks)

DROOLING (SIALORRHEA)

Glycopyrrolate (pill) - Reduces saliva; well tolerated; May cause dry mouth, constipation
Scopolamine Patch (transdermal) - 72-hour patch for saliva control; helpful if patient cannot swallow meds; can cause dry mouth or confusion
Amitriptyline (pill) - reduces drooling + helps sleep + helps PBA; sedating; strong dry-mouth effect
Atropine Drops (sublingual) - Quick, short-acting saliva reduction; can cause dryness; may be difficult for them to squeeze out

JAW CLENCHING / SPASTICITY / MUSCLE ISSUES

Baclofen (oral) - reduces spasms/spasticity; sleepiness + weakness possible
Tizanidine (oral) - Spasticity management; can cause low BP, sedation
Benzodiazepines (clonazepam, diazepam) - Jaw spasms, anxiety, spasticity; sedating; fall risk
Botulinum Toxin (Botox) - For jaw-closing spasms or salivary glands; may temporarily affect swallowing

PSEUDOBULBAR AFFECT (PBA)

Nuedexta - First-line for uncontrolled laughing/crying; generally well tolerated
SSRIs (sertraline, fluoxetine, fluvoxamine) - Useful when depression/anxiety also present
Tricyclics (amitriptyline, nortriptyline) - Helps PBA + drooling + insomnia

PAIN MANAGEMENT

Gabapentin / Pregabalin - Neuropathic pain; sedating
NSAIDs (Ibuprofen, Naproxen) - Musculoskeletal pain; stomach irritation possible
Tramadol - Moderate pain; sedation + constipation

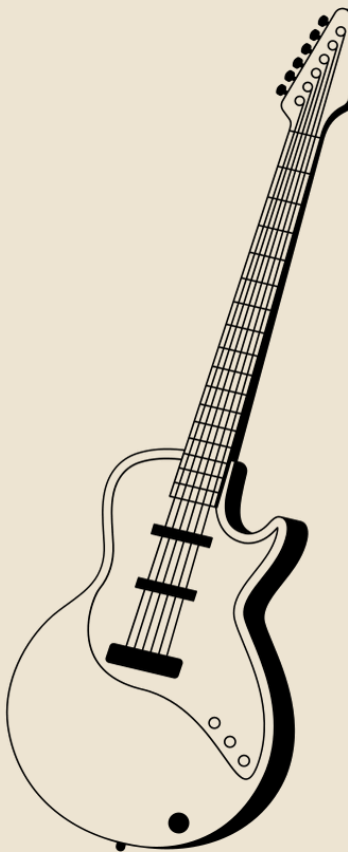
MOOD / ANXIETY / APPETITE

SSRIs (Zoloft, Prozac, Celexa, Paxil) - Depression, anxiety; possible nausea, sleep changes
SNRIs (Cymbalta, Effexor); Depression + neuropathic pain
Mirtazapine (Remeron) - Boosts appetite + helps insomnia; very sedating
Bupropion (Wellbutrin) - Depression + low energy; not great for anxiety

GI / APPETITE / REFLUX

Metoclopramide (Reglan) - Early satiety; moves stomach contents; watch for restlessness or movement side-effects
Acid Reducers (Pepcid, Zantac, Nexium) - Reflux contributing to cough, low appetite

Patients often take vitamin pills which are typically large. These vitamin pills can usually be consumed in a different form, if swallowing is a concern.



No One is an Island

HER ALS STORY

Leah's ALS journey began in 2019, when she finally got an answer for why her normally active body wasn't keeping up. As a young woman facing a life-changing diagnosis, she wanted to find others her age going through similar experiences. Two years later, she felt ready to channel that into advocacy and connected with I AM ALS and advocate Lori Andre. With their support, she launched #InHerALSShoes to push back against the stereotype that ALS only affects older white men. The momentum from that project led her to create Her ALS Story in 2021–a space for young women with ALS to support each other and speak up in a powerful, united way. Today, the group has more than 100 members around the world and works with organizations like I AM ALS and Project ALS to strengthen existing efforts and highlight what it's really like to be a young woman living with ALS.

Read about Women's ALS Journeys

Amazon Storefront with Organized Folders

Thank you for reading!
For any questions, updates,
edits, or error corrections, feel
free to reach out at
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References