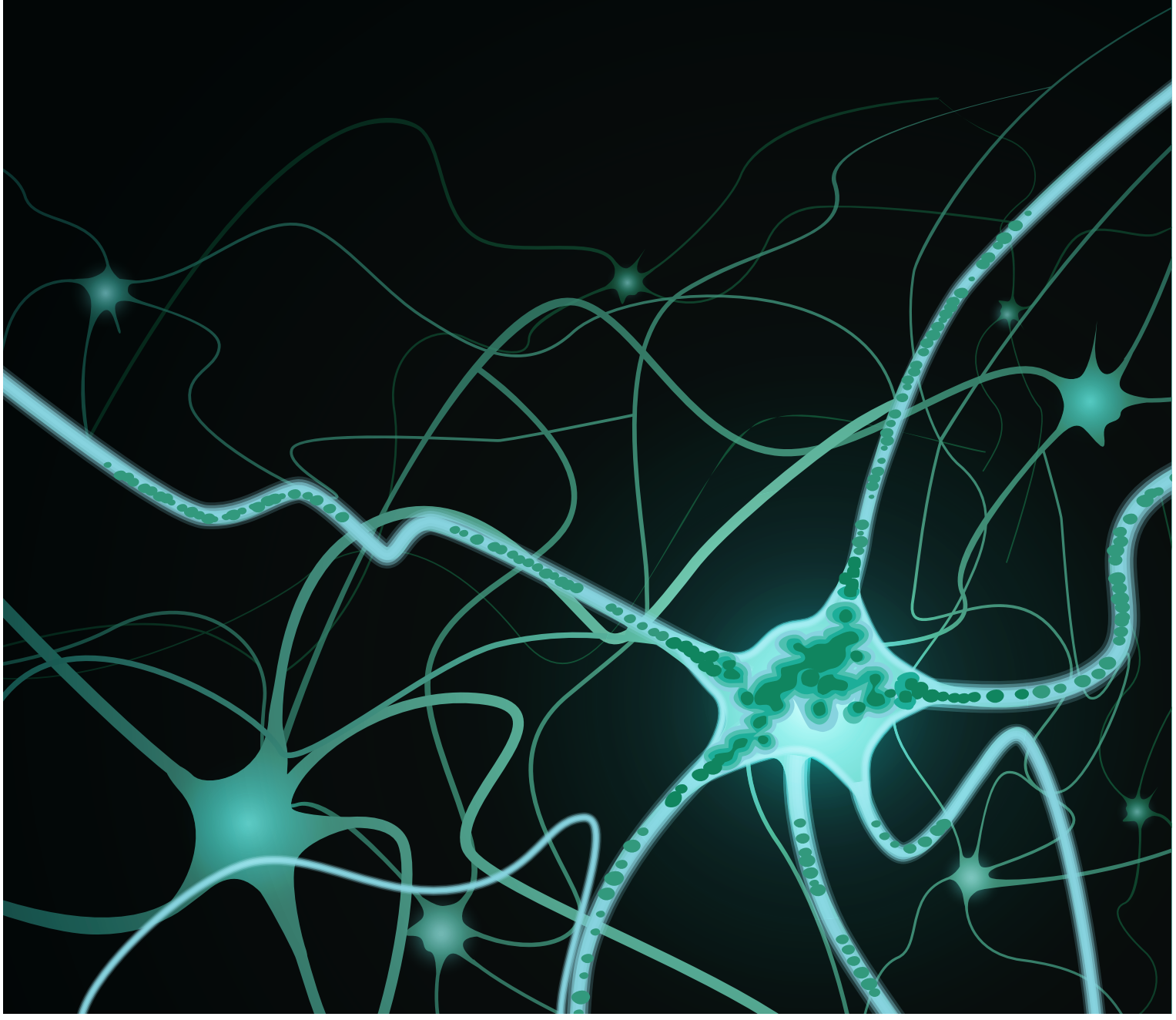
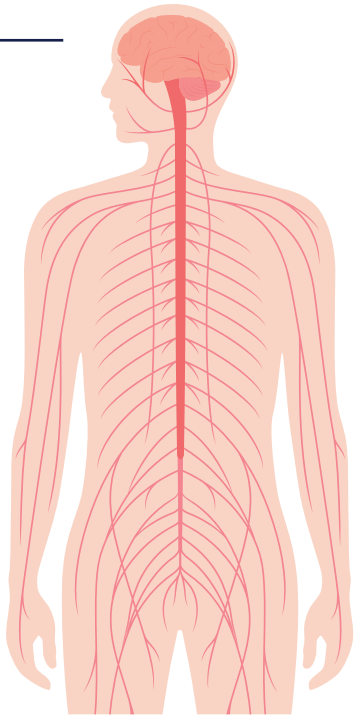
AUSTIN
NEUROMUSCULAR
CENTER

A Guide to Understanding and Living with Amyotrophic Lateral Sclerosis (ALS)



What is ALS?

Amyotrophic Lateral Sclerosis (ALS) is a progressive, neurodegenerative disease and is fatal. ALS attacks nerve cells in the brain and spinal cord that control muscle movement. ALS robs you of your ability to walk, talk, and eventually breathe. It is always fatal.



BREAKING IT DOWN

a-myo-tro-phic lateral sclerosis | noun | Greek

definition: no muscle nourishment

"a" ..... means no or negative

"myo" ..... refers to muscle

"trophic" ..... means nourishment

"lateral" ..... is the area in the spine where the brain tells the muscles what to do

"sclerosis" ..... is hardening; as the disease progresses, the lateral areas harden and the signals stop

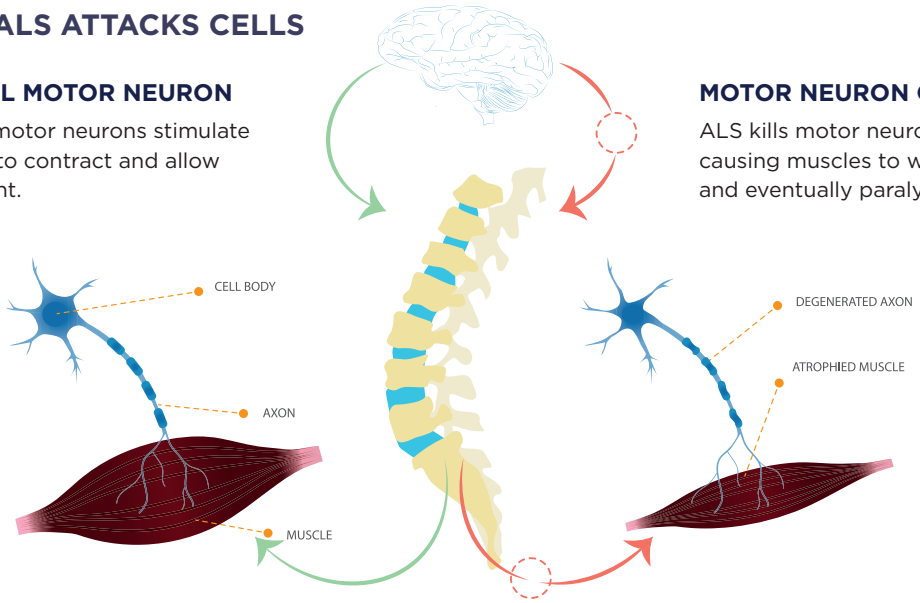
HOW ALS ATTACKS CELLS

NORMAL MOTOR NEURON

Healthy motor neurons stimulate muscles to contract and allow movement.

MOTOR NEURON OF ALS

ALS kills motor neurons, causing muscles to weaken and eventually paralyze.

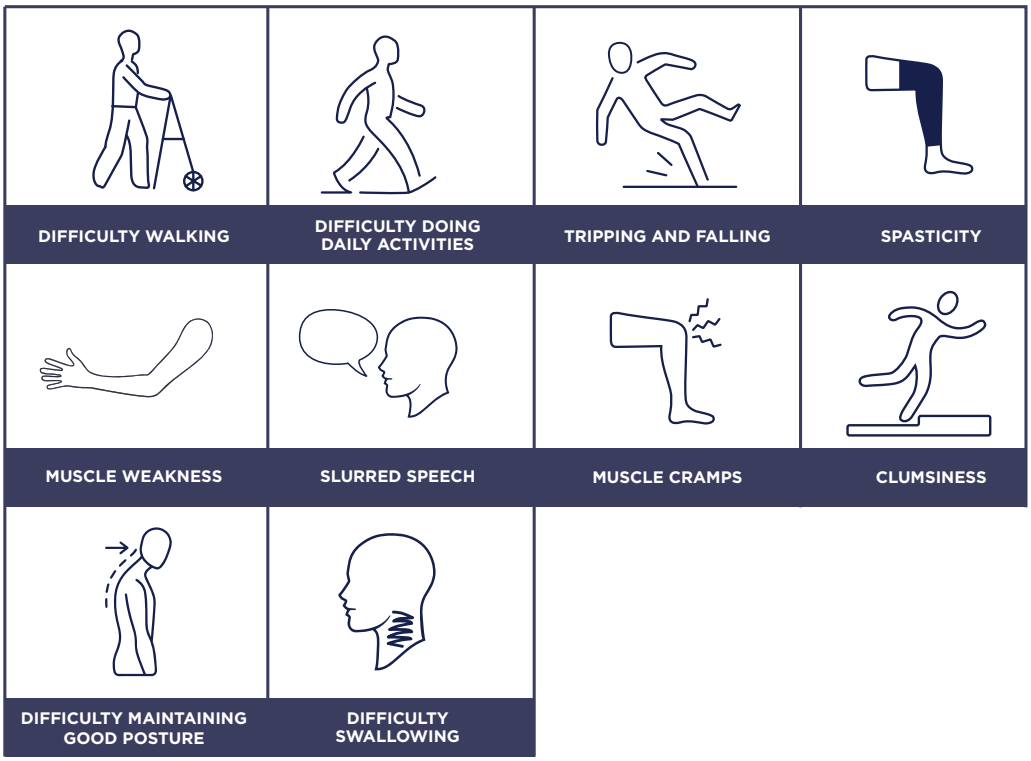


PROGRESSION AND DIAGNOSIS OF ALS

ALS leads to a progressive loss of muscle control, and gradually prohibits the ability to:

- Speak
- Swallow
- Walk
- Grasp objects
- Move
- Breathe

SYMPTOMS OF ALS



ALS patients will have even more difficulty in breathing, chewing, speaking and swallowing as the disease progresses and their condition deteriorates. Eventually the patient may be unable to breathe on their own, requiring the use of a ventilator.

Every patient's experience with ALS is a little different. See the following pages for resources and what to do about your symptoms.

FACTS ABOUT ALS

5,000+ people are diagnosed per year

Only **4 drugs** are currently approved by the U.S. FDA to treat ALS (Riluzole, Nuedexta, Radicava, and Tiglutik)

2–5 years is the average life expectancy

$2 billion is the estimated cost to develop a drug to slow or stop the progression of ALS

10% of cases are inherited through a mutated gene

$250,000 is the estimated out-of-pocket cost for caring for a person with ALS

90% of cases occur without a family history

There is **no cure** for ALS

Every **90 minutes** someone is diagnosed or someone passes away from ALS

Veterans are more likely to get ALS. ALS impacts veterans regardless of the branch of service they served in and affects those who served in both peacetime and war.

FDA APPROVED MEDICATIONS

Riluzole (Rilutek): blocks release of glutamate (glutamate injures the nerve cells)

- Shown to extend life an average of three months
- Liquid form available
- Side effects: abdominal pain, nausea

Edaravone (Radicava): free radical scavenger; IV infusion

- Only for early ALS; no affect on mortality but showed a 2 point increase on the ALS-FRS (symptom questionnaire)

Radicava ORS: newly approved and same as above but in oral form

- Side effects: headache, gait disturbance

Relyvrio: newly approved, oral packet; combination of sodium phenylbutrate and taurursodiul

- In clinical trials, shown to increase the ALS-FRS score by 2.3 points and extend life 4.8 months
- Side effects: nausea, diarrhea, upper respiratory infection

Nuedexta: used for Pseudobulbar Affect (PBA) (inappropriate laughing and crying); combination of dextromethorphan and quinidine

- Side effects: diarrhea, dizziness

We have several clinical trials at our clinic. Ask us for details on enrolling. We would be happy to discuss trials that are elsewhere.

MANAGING YOUR ALS SYMPTOMS

MOBILITY

You may start to have difficulty walking, which can lead to falls. You may also experience spasticity or stiffness in your arms or legs, making it more difficult to do certain tasks. When you begin having difficulty walking, you may need braces. Eventually, you may need assistive devices like a cane, walker, scooter, or wheelchair.

Ankle Foot Orthosis (AFO) braces are a good way to prevent falls. We recommend getting fitted by and purchasing from Hanger Clinic. There are several locations in Austin. Below is the address to their central location:

Hanger Clinic
7020 Easy Wind Dr, Suite 130
Austin, TX 78752
Phone: (512) 377-2323

For spasticity, we may try different medications like dalfampridine, gabapentin, or muscle relaxers.

It's important to keep moving and know how to work with your limitations. Physical therapists that are specialized in neuromuscular conditions can be helpful.

EXERCISE RECOMMENDATIONS

Exercise is important for those with ALS for flexibility, maintaining strength, and mobility, but not all exercises are helpful. Since ALS affects people differently, there isn't one exercise program for all. Discuss with your physical and occupational therapist on specific recommendations.

- Try to stick with range of motion and stretching exercises to prevent further damage to the muscle.
- If your muscles are weak to the point where it takes a lot of effort to move them, trying to lift additional weight may be harmful. Also, if you feel overly weak after exercise then you may be doing too much.
- Stretching improves flexibility and can help correct imbalances in the muscles and tendons on each side of a joint.
- Stretching can be active, meaning you do the stretching yourself, or passive meaning someone moves your limbs for you.
- You can use strengthening exercises to preserve muscle strength. Choose a weight with which you can perform 20 repetitions, but only do 10. This would only be done with limbs that have full range of motion. You don't want to overwork the muscles.
- Some people find pool therapy helpful as there is buoyancy to help move your limbs and less danger in falling.
- Establishing an exercise program with physical and occupational therapy is helpful in that they can modify recommendations as things change.

DYSPHAGIA: DIFFICULTY SWALLOWING

You may have difficulty swallowing either by choking on food and/or water, or your jaw becoming fatigued from chewing. You may have to change the consistency of your food (i.e., thicker foods). It is vital that you not lose weight. We recommend a feeding tube (PEG) if you lose 10 percent or more of your body weight after diagnosis.

Symptoms of dysphagia include:

- Choking on food or drink
- Coughing during or after swallowing
- Coughing or vomiting up food
- Having a weak, soft voice
- Aspirating (getting food or liquid in your lungs)
- Excessive saliva or drooling
- Difficulty chewing
- Trouble moving food to the back of your mouth
- Food sticking in your throat

DYSARTHRIA: DIFFICULTY SPEAKING

Your speech may change by sounding slurred or spastic (slow, nasal, strained).

Speech therapy can help with both dysphagia and dysarthria.

DIET AND NUTRITION

It is important to eat a well-balanced diet to provide energy, help fight infection, and prevent weight loss. Losing weight and malnutrition can cause worsening muscle weakness and progression of ALS. Your energy demand is actually higher than someone without ALS.

What and how you eat may need to be modified if you are having chewing or swallowing issues as well. Try to get adequate fluids because constipation is common with ALS. Try not to lose weight, even if you feel you are overweight.

Resources

Maintaining good nutrition with ALS

FOOD GROUPS	DAILY SERVINGS	EXAMPLES
Meats and alternatives	Three or more	• 2 oz. beef, poultry, fish • 2 eggs • 2 oz. cheese • 1 cup cooked beans
Dairy	Two or more	• 8 oz. milk • 1 cup yogurt (Greek yogurt has twice the protein of regular yogurt)
Fruits and vegetables	Four to six	• ½ cup canned fruit • 1 piece of fresh fruit • ½ cup cooked vegetables • 1 cup fresh vegetables
Grains and starches	Six to eleven	• 1 slice of bread • 1 cup ready-to-eat cereal • ½ cup cooked cereal • ½ cup pasta or rice • ½ cup mashed potatoes
Fats	Four to seven	• 1 tsp. butter or olive oil • 1 tbsp. cream cheese • 1/8 avocado • 1 tsp. mayo or salad dressing
Desserts	One or two daily to maintain weight	• rice/bread pudding • chocolate/vanilla pudding • ice cream • frozen yogurt • smoothie/shake

Note: A person with swallowing difficulties may need to modify food consistencies with the guidance of a Speech/Language Pathologist (SLP).

DYSPNEA: SHORTNESS OF BREATH

You may start to have shortness of breath during activities you do every day. Research has shown that having adequate breathing support and maintaining weight are the two most important factors in extending life with ALS.

We recommend non-invasive ventilation (NIV) when your breathing values are less than 50 percent or you have difficulty breathing while lying flat (orthopnea). When you get to this point, we will send a referral to a pulmonology or respiratory clinic for you to be fitted for and educated about NIV.

SIALORRHEA: DROOLING

If you are having swallowing or speech problems, you may begin to notice drooling or difficulty managing your secretions. There are a few medications that can be used to help, including scopolamine, glycopyrrolate, nortriptyline, atropine drops, and in some cases, Botox injections.

PSEUDOBULBAR AFFECT: INAPPROPRIATE CRYING AND LAUGHING

Some people with ALS may experience a condition called Pseudobulbar Affect that's characterized by episodes of sudden uncontrollable and inappropriate laughing or crying. People with bulbar (affecting muscles of the throat) ALS and/or with genetic mutation of C9ORF72 may be at higher risk of developing PBA. There is an FDA approved medication, Neudexta for PBA.

FRONTOTEMPORAL DEMENTIA (FTD)

Approximately 15% of ALS patients meet criteria for FTD. There may be a link between FTD and having C9ORF72 genetic mutation as well. People may experience changes in personality, dietary preferences, word-finding difficulty, as well as cognitive and other behavioral changes. Diagnosis is made by clinical testing and imaging. Treatment is usually supportive and focused on health maintenance and safety.

CRAMPS AND PAIN

Pain may arise from a variety of causes like limited mobility, cramps, spasticity (muscle stiffness), and co-morbid conditions. Some people may have sensory symptoms like numbness and tingling. Symptoms can be treated conservatively with methods such as using heat and cool packs, massage, repositioning, and over the counter medications. If pain and cramps are severe, neuropathic pain medications like gabapentin, pregabalin, or nortriptyline can be used. The patient may also benefit from a reduction of saliva, which is a side effect of nortriptyline. Medications for cramps include magnesium supplements, gabapentin, medical cannabis, and muscle relaxants.

THE USE OF ANESTHESIA WITH ALS

Anesthesia can be risky with ALS because of diaphragm and other respiratory muscle weakness. There is also risk of aspiration with weak swallowing muscles. Short acting agents are preferred and epidural anesthesia has been used as well. There is no ideal anesthesia, and the risk increases as the disease progresses. If needing surgery (ex: PEG), sooner is better than later.

ALS ASSOCIATION
The ALS Medicare Resource Line

The ALS Association partnered with Patient Advocate Foundation to offer the ALS Medicare Resource Line. This program offers free and confidential navigational assistance with financial and practical challenges that impact your ability to access healthcare.

Living with ALS can be overwhelming, and can complicate daily life and cause extreme stress for patients and families. The Patient Advocate Foundation can help patients with:

ACCESS TO CARE

- Getting prescribed medical treatment and services
- Understanding what your health insurance covers and your options if you don't have insurance

PAYING FOR TREATMENT

- Getting approvals and payments from health insurance
- Appealing insurance denials
- Applying to insurance programs, like Medicaid, Medicare, and the Health Insurance Marketplace
- Applying to programs that help pay for co-pays and insurance premiums
- Applying for free or low-cost healthcare and medicine programs
- Getting discounts or setting up payment plans

PAYING FOR LIVING EXPENSES

- Applying for programs that can help pay for things like food, rent, utilities, and transportation
- Applying for Social Security Disability Insurance (SSDI)

EMPLOYMENT

- Helping you use your employee benefits, like health insurance and sick leave
- Understanding laws like the Americans with Disabilities Act (ADA) and the Family and Medical Leave Act (FMLA)
- Applying for disability insurance to help with income if you can't work; and helping you challenge disability denials, if needed

Request help or learn more about available services at als.pafcareline.org or call (844) 244-1306

RECOMMENDED PROVIDERS

Ankle Foot Orthosis Braces

Hanger Clinic
7020 Easy Wind Dr, Suite 130
Austin, TX 78752
Phone: (512) 377-2323

Dysarthria

Moira Gallagher, SLP

Dyspnea

Respiratory Quality Services (RQS)
12830 Murphy Road
Stafford, TX 77477
(713) 349-9008

RQS Austin/Round Rock
Bronwen Surber, CRT
Manager, Austin/Round Rock
(512) 497-9885
bronwen@rqsbreathe.com

Pulmonology and Critical Care Associates of Austin
Dr. Michael Shapiro
1305 W 34th Street, Suite 400
Austin, TX 78705
(512) 459-6599
samantha@pccca.com

Physical Therapy

Georgetown Living
Spero Rehab

Resources

Everyday Life with ALS
(chapter 8 has exercises)

Range of Motion Exercises

Exercise and Fitness