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Also known as ALS or Lou Gehrig's disease, amyotrophic lateral sclerosis is a rare neurological disease involving the breakdown, and eventual death, of neurons that control voluntary muscles. The brain loses the ability to initiate and control movement, often resulting in an inability to eat, speak, move, and even breathe. Approximately 5,000 people in the US are diagnosed with this condition per year, according to the ALS Foundation. There are treatments for ALS, but there is no cure. Loss of coordination is one of the first warning signs of ALS. Reduced hand-eye coordination may start slowly, with the individual noticing relatively minor issues, such as the ability to grasp a hairbrush. Over time, the number of episodes increases in frequency, and in some cases coordination issues last for months before other symptoms begin to appear. An early warning sign of ALS is cramps or muscle spasms, especially when they develop with other symptoms. Cramping, twitching, and atrophy of the muscles occur when spinal and brain stem motor neurons deteriorate. This particular symptom typically begins after the loss of hand-eye coordination and can last through many of the disease's stages. ALS almost immediately begins to wreak havoc on the nervous system. Motor neurons that tell muscles to move begin to die, and as a result, individuals lose control of and strength in their muscles. The degeneration of neurons leads to loss of muscle mass, too. Due to the lack of muscle mass, people with ALS often require wheelchairs even before they lose the ability to walk. Laryngeal dysfunction can occur when the loss of neurons affects the bulbar nerves. Individuals with ALS often experience changes in voice pitch, usually resulting in speaking at a lower register. This common symptom typically coincides with hand-eye coordination issues. Slurring of speech happens for a different reason than the change in vocal pitch. This symptom occurs when the person is no longer capable of properly moving muscles involved with speech, including the lips and tongue. This is generally referred to as dysarthria, the inability to speak correctly due to loss of motor function. The individual affected may find it difficult to pronounce words properly. Some people with ALS experience significant mood swings that can result in uncontrollable laughter or crying, known as the pseudobulbar affect. Although it is not completely understood why this occurs, it is believed that when upper motor neurons, located in the cerebral cortex and brain stem, deteriorate, the disrupted neural pathways result in a deficiency of inhibition control. Emotions can come on suddenly, without warning, and often without triggers, lasting only a few moments or up to an hour. Prescription medications can help control the onset and amplification of emotions. Problematic breathing does not occur immediately in all people with ALS, but most experience it eventually. As muscles deteriorate, those responsible for bringing air in and out of the lungs are also affected. An individual may first develop shortness of breath and an inability to breathe in deeply. This can progress to collapsed lungs and a continuous need for a breathing machine. Trouble walking is a common ALS symptom usually seen in later stages of the disease. Initially, an individual may experience weakness in the legs, which gradually worsens as the disease continues to cause muscle degradation. There are many possible causes of weakness in the legs; however, most people with ALS experience other symptoms before this one peaks. Swallowing problems are another late symptom of ALS. The issue can make it difficult to eat and drink, which makes choking a constant danger as it becomes difficult even to swallow saliva. This symptom is caused by the lack of control of the muscles used when swallowing. Primary doctors may work with dietitians to devise meal plans that ensure individuals with ALS continue to get proper nutrition. People with advanced ALS often experience an inability to control the muscles in the neck, resulting in a condition called dropped head syndrome. The muscles at the back of the neck weaken, which is why people with ALS often require neck braces. Medically Reviewed by Christopher Melinosky, MD on November 19, 2021 Amyotrophic lateral sclerosis, or ALS, is a disease that attacks the nerve cells in your brain and spinal cord. There is no known cure. But doctors do have treatments and therapies that can slow down or ease symptoms in you or a loved one. Researchers continue to study ALS, hoping to learn more about its causes and possible new treatments. It's commonly known as Lou Gehrig's disease, after the baseball player whose diagnosis and eventual death brought wide public attention to the illness. This condition kills the nerves that control motion in your body. As those nerves die, you lose control of your muscles. As the disease worsens, you lose the ability to walk, speak, swallow, and eventually, to breathe. About 1 person in 25,000 will be diagnosed with ALS. Most of them die within 2 to 5 years of being diagnosed, usually because of respiratory failure. However, a small group, about 5% of those with ALS, have been able to survive for 20 years or more. There are two medications which have proven helpful in slowing the progression of ALS and extending the life of those who have been diagnosed with the disease. While they have been shown to push back the time when you'll need mechanical help to breathe, they can't fix damage already done. Edaravone (Radicava), administered through IV, it is an antioxidant that can prevent damage to nerve cells from toxic substances called free radicals. But it is unclear how it works to slow the physical progression of patients with ALS. The most common side effects include bruising, unsteady gait, and headache. Riluzole (Rilutek): taken orally, it helps reduce damage to your motor nerves by reducing the amount of glutamate in your system. (Glutamate carries chemical messages to your nerves. Too much of it can damage the cells). The most common side effects include gastric distress, dizziness and bruising. Pain relievers or muscle relaxants such as baclofen (Gablofen, Kemstro, Lioresal) or diazepam (Diasat, Valium) can help ease cramps. A variety of medications can lower how much saliva you make. It often builds up in your mouth as swallowing becomes more difficult. One of the most common medications is glycopyrrolate (Robinul). Doctors may prescribe drugs to help ease other symptoms of ALS, which may include: Constipation Depression Outbursts of laughter or crying Lack of sleep Fatigue Most treatments for ALS involve managing the symptoms of the disease as it worsens. Some of them include: Physical therapy and exercise: These keep your muscles strong and working as long as possible. Hot tub and whirlpool baths: These can ease your muscle spasms or cramps. Dietary counseling: This becomes really important when swallowing becomes a problem. Speech therapy: Specialists can help you learn ways to make your speech more clear when you talk or other methods of communicating, such as writing with pen and paper or an alphabet board. Occupational therapy: This can help you find ways to dress, bathe, and groom. A therapist can help you set up your home so it's easier for you or a loved one to move around. There are also a variety of tools and mechanical devices that can help if you have ALS: Splints, reach extenders, and grab-bars: They can help you get around as the disease progresses. Canes, walkers, and wheelchairs: They can help you stay mobile even as your ability to walk fades. Computerized voice synthesizers: These are available when you lose the ability to speak. In the final stages of the illness, only about a quarter of people can talk clearly enough to be understood. Respirator: It may be needed in the late stage of the disease to help you breathe. A doctor will have to insert a breathing tube directly into your windpipe. This is called a tracheostomy. Feeding tube: As swallowing becomes harder, you might also need a doctor to insert a feeding tube into your stomach. This tube is called a PEG tube (percutaneous endoscopic gastrostomy). © 2021 WebMD, LLC. All rights reserved. View privacy policy and trust info Medically Reviewed by Arefa Cassoobhoy, MD, MPH on November 20, 2020 Many things about amyotrophic lateral sclerosis (ALS), also known as Lou Gehrig's disease, remain unclear. Without knowing exactly what causes ALS, it's hard to tell why some people get the disease while others don't. Researchers do have some possible ideas, however. ALS disrupts your motor neurons. These are nerve cells that control important muscle activities, including breathing, talking, swallowing, and walking. Over time, the loss of muscle control becomes worse. There is no cure for ALS, although research is ongoing. There are no preventive steps either. It's rare, affecting about 5.2 people per 100,000 in the U.S. population, according to the National ALS Registry. Because of the seemingly random nature of the condition, it's hard for researchers to pinpoint who might have a greater chance of getting it. Doctors have learned some things from people who have this condition. ● Sex: About 60% of people with ALS are male. ● Race: 93% of people with it are white. ● Aging: Although the disease can strike at any age, symptoms most commonly develop between the ages of 55 and 75. You can get it earlier, though having it before 30 is very rare. ● Family history: A small percentage of ALS cases are passed down from family. There are two main kinds, depending on whether the disease runs in your family. ● Sporadic: This makes up 90% to 95% of all ALS cases, as it occurs in people who have no known family history of the disease nor any clear things that would make them more likely to get it. Other family members are not expected to be at risk for inheriting ALS in sporadic cases. ● Familial: In about 5% to 10% of cases, ALS runs in the family. If you have a familial ALS, there is a 50% chance that your children will get it as well. Scientists are looking into whether genetics, things in the environment, or a combination of both cause ALS. Some theories suggest people who might already be genetically at risk for ALS get the disease after some kind of contact with an outside "trigger" in their environment, such as being around a toxin. Scientists have found over a dozen mutations in genes that have ties to ALS, but the two major ones are C9orf72 and SOD1 genes. C9orf72 gene: Mutations in the gene known as C9orf72 have been found in about a third of all familial cases and a small percentage of sporadic ones. Scientists have also found that this defect on the C9orf72 gene is tied to what's called "frontotemporal dementia (FTD)," an uncommon form of dementia. Some researchers think that ALS and some forms of FTD are related. SOD1 gene: Mutations on this gene appear in about 20% of familial cases and 1% to 5% of sporadic ones. It's unclear how the mutations lead to ALS. Research has found that proteins from a mutated SOD1 gene can become toxic. Scientists are also looking at whether things in the environment such as chemicals and other agents can raise your chances of getting ALS. But it's been hard for them to prove anything specific so far. Some things they are looking into: Smoking: Smoking is believed to be the only probable factor that may raise your chances for ALS. But this may be true mainly for women, especially those after menopause. This link is controversial among doctors. Contact with toxins: Lead and other chemicals may be linked to ALS, but no single agent has been consistently found to be a cause. Military service: Studies have found that military veterans, especially those deployed during the Gulf War in 1991, have a greater chance of ALS. The exact causes remain unclear, but may include contact with chemicals or metals, injuries, infections, or the intense physical activity needed to serve. Those who were in the Gulf War are more likely to get ALS compared with other veterans. Intense activity: The most famous person to have ALS was Lou Gehrig, the baseball player who died from it. Studies have shown a higher chance among athletes, who are very active. But the studies have been small, so it's too early to say that being an athlete means you have a greater chance of getting the condition. Your work: Several lines of work – including sports, cockpit, construction, farm, hairdressing, lab, veterinary, and welding, among many others – have been reported to carry a higher chance of ALS. These jobs often involve some kind of contact with pesticides, metals, and chemicals. But the common, underlying risk has not been found. Where you live: Clusters of ALS cases have been reported on the Pacific island of Guam and in the Kii Peninsula in Japan, which have rates 50 to 100 times higher than other parts of the world. Such clusters have also been reported in South Dakota and Italy. © 2020 WebMD, LLC. All rights reserved. View privacy policy and trust info





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