TIPS

Practical advice for nurses and caregivers dealing with ALS in homecare

Those working in homecare do not have to deal with Amyotrophic Lateral Sclerosis (ALS) all that often in their careers. For that reason, they have very limited knowledge or experience with this drastic disease. That may be understandable, however, it can also cause a problem when someone suffering from ALS needs homecare. This folder provide information about ALS for nurses and caregivers in homecare and also elaborates on the special attention someone with ALS requires. The information about ALS is for a large part the same as for the rapidly evolving form of progressive spinal muscular atrophy (PSMA). The symptoms that occur in this rapid variation of PSMA are similar to those of ALS. ALS is a rare yet complex disease of the central nervous system and muscles. The care someone with ALS needs is similar to the care people with other complex diseases need, however, some treatments and symptoms require special attention.

- The first part of the folder contains brief information about the disease.
- The second part provides some practical advice.

In general

Be clear about the coordination.
In ALS it is of great importance that one nurse will act as the care coordinator and will be responsible for the proper coordination of the home care to the situation of the patient with ALS. The nurse has a main role in the caregiving. It is important that everyone involved knows who is primarily responsible.

Keep up-to-date with the evolution of the disease
Symptoms can change very quickly. Make sure that with every visit you keep track of what changed in the meanwhile. Read the care file very carefully and stay informed by the patient or caregiver about the last developments and keep the file updated.

Be aware of personal circumstances and needs
The progression of ALS is partly predictable but not every person with ALS has the same symptoms in the same order. The rate at which the disease progresses may vary from person to person. Always pay attention to the personal circumstances and the needs of the person receiving the care.

1. What is ALS?

Amyotrophic lateral sclerosis (ALS) is a disorder of the nervous central system which worsens rapidly and gradually causes more muscles to stiffen and weaken. That has profound implications for moving, speaking, swallowing and breathing. ALS occurs in four to six people on a scale of hundred thousand, mostly between the ages of forty and sixty. In more than ninety percent of ALS cases ALS is not inherited. Since the cause of the disease is unknown, the treatment consist mostly of fighting the symptoms.

Course of the disease

The effects of ALS are getting worse, without the hope of a cure. The disease progresses at a fast rate. Within the course of three years on average, the respiratory muscles have weakened so that independent breathing is no longer possible, resulting in death. About twenty percent of people with ALS live for only five more years after the first symptoms.

Two forms
The sequence in which the muscles are affected in ALS can vary. In overall, a distinction is made between a spinal and a bulbar form of ALS.

The spinal form starts in the bone marrow and leads to loss of strength in arms, legs and torso. That causes movement problems which often start with tripping, falling or the dropping things. Due to the increasing loss of muscle strength, over a period of time all kinds of operations will become more difficult and even impossible. In time, the strength of the chewing and swallowing muscles will also weaken, which will cause problems with swallowing, chewing and speaking. The spinal form presents in about sixty-five percent of people with ALS.

Early onset, the bulbar form causes especially weakness in the muscles of the mouth and throat area, making talking, eating and drinking significantly difficult. In this form, sometimes even compulsive crying, laughing or yawning may occur. Over time, the symptoms will also appear in the arms and legs. In about thirty-five percent of people with ALS, the disease begins with bulbar symptoms.

Treatment and medication
The cause of ALS is unknown. ALS causes nerve cells to

die off, making it difficult for signals from the brains to reach the muscles. A good rehabilitation medical treatment contributes significantly to the quality of life. In Belgium, rehabilitation centers exist, however, they are not specifically ALS oriented. In a matter of time, a care center will be established where the same care will be provided specifically for ALS patients. At the moment, various assistance is coordinated by the Neuromuscular Reference centers. (NMRC).
As long as the exact cause of ALS is unknown, the disease cannot be treated. However, the development of the disease may be delayed by three to six months with the drug riluzole, known under the brand name Rilutek. Up to now, it is the only drug for ALS of which the effect has been proven. Riluzole is prescribed by the neurologist. A person with ALS also can also receive medication for symptoms such as compulsive laughing, crying or yawning and muscle spasms.

Important symptoms
As mentioned above, the form of ALS determines the part of the body in which the symptoms will first occur. That means that not everyone with ALS immediately start experiencing speech and swallowing problems or issues in the arm or leg muscles. Eventually it almost always creates weakness in all muscles including respiratory muscles. The extent and severity of the symptoms and the speed in which they worsen vary in each individual case.

Muscles
Beginning muscle weakness can lead to tripping, experiencing difficulty in turning a wrench or an unclear pronunciation. If the muscle loss takes longer, atrophy of the muscles occur; they become thinner. Since the muscle function in arms, legs, mouth and throat further deteriorates, it becomes increasingly difficult to move, eat or speak independently. Eventually a person with ALS will become dependent for all activities of daily life on the help of others.

Another phenomenon in ALS is that small movements often may cause unusual cramps in the muscles of the hands, feet and calves and in the muscles of the mouth. The muscles stiffen and can cause spasticity in action. Because of the disorder in the nervous system, people with ALS suffer from fasciculations; small muscle movements under the skin that sometimes can be very annoying.

Swallowing
Due to muscle weakness in the mouth and throat area, swallowing becomes increasingly difficult, which increases the risk of choking. By choking, food can get into the lungs which can cause a pneumonia. People with ALS who eat less due to their difficulty in swallowing food, often lose weight. For that reason, in many cases a PEG tube is placed in an early stage, so that the person with ALS can switch to tube feeding, when swallowing becomes difficult. Tube feeding is usually combined for a certain period with solid food before someone completely switches to tube feeding. A nutrition team of the hospital or the ALS rehabilitation team can offer guidance for swallowing difficulties. It is equally important for the home care nurse to have the proper expertise in dealing with swallowing issues. Because ALS patients cannot swallow all of their saliva, they suffer from excessive salivation. In other words, they are going to drool, which may be experienced as annoying and lead to social isolation.

Speech
People with a bulbar form of ALS often have speech problems and may ultimately not be understandable anymore. They become dependent on other forms of communication. Although, people often become proficient in the use of such speech replacing devices, they remain dependent on the patience of their interlocutors to communicate with them.

Breathing
People with ALS may also show symptoms as a result of the weakening of the respiratory muscles. This is a gradual process. The first symptoms often occur at night because breathing during sleep is less efficient and waste is not properly exhaled. The accumulation of waste products in the body is called hypoventilation. Over time, this phenomenon also occurs during the day. Symptoms that may occur are therefore shortness of breath, restless sleeping and dreaming, inability to lie flat, morning headaches and drowsiness during the day.

An oxygen deficiency does not yet occur in a beginning hypoventilation. That may arise when aside from a hypoventilation, respiratory infections occur. In that case, it is important to contact a doctor so that he can decide whether or not to administer oxygen. To treat the symptoms of hypoventilation, a Neuromuscular Reference Center can provide a form of respiratory support. Not everyone chooses for this option or gets used to it. It is often necessary for a patient to be admitted for the parameter settings of the control equipment. The respiratory support often starts at night with breathing through a cap placed on the nose and mouth. This leads to an alleviation of the symptoms and slows down the disease progression a little.

When respiratory support also becomes necessary during the day, the patients has the possibility to switch to chronic invasive ventilation via a tracheostomy. However, the ALS Center and the Neuromuscular Reference centers are not in favor of invasive ventilation. This form of breathing requires the continuous presence of someone who can drain off the saliva if necessary. Although this option often implies an extra burden for the caregiver, the patients and their loved ones may have their reasons to opt for this solution.

Thinking and feeling

A person with ALS continues in most cases to feel and think normally. He or she may experience difficulties with expressing themselves in speech or with gestures. Therefore, it is sometimes said that a person with ALS becomes trapped in his or her own body. In about five to ten percent of patients with ALS, frontotemporal dementia occurs.

Typical for this form of dementia are behavioral changes associated with insufficient insight in the disease and emotional numbness. ALS has no effect on the heart muscle, the eye muscles and the sphincter muscles. A person with ALS continues to have good vision, hearing and smell and is usually not incontinent. Sexual functions also remain intact, touches are still felt and it is still possible to have an erection or orgasm. At the same time intimate physical contact becomes more and more a one-way street as a result of the symptoms of paralysis. This development often requires adaptation to different roles in the relationship. Often, the partner will have to take over tasks and increasingly fulfil a caring roll. The person with ALS may begin to struggle if the equality in the relationship is at risk of being lost.

Perception of the client and his/her relatives

Most people experience the diagnosis of ALS like a bolt from the blue. They are often still in the middle of their life, with a busy job, a lot of extracurricular activities and sometimes even young children. Information about the rapid and fatal course of the disease may have a devastating effect, bringing along fear, sadness, anger and insecurity. After the diagnoses, it is unforeseeable what the disease will personally imply for someone. ALS forces people to continually adapt to new situations, make profound choices and adjust their expectations. After a while, most people find a way to cope with it in their own way.

Fear

Many people diagnosed with ALS, are afraid that they will die by asphyxiation. Tightness of the chest therefore often leads to panic. Research shows that people with ALS do not choke, but that over ninety percent dies quietly. Due to the accumulation of carbon dioxide, they become increasingly dazed and finally pass away quietly. Tightness of the chest can be caused due to choking, difficulties with coughing up slime or a pneumonia.

Loss

As the disease progresses, people with ALS increasingly need to give up their independence of physical functions. This can cause feelings of sadness and anger. Having to learn to live with ALS is therefore sometimes described as a continuous process of mourning.

In the bulbar form it is a frustrating experience to lose speech or the inability to eat. It is also a significant step to undertake in switching to a PEG tube. It also requires a lot adaptability to learn to communicate differently. Moreover, it is not easy to cope with other signs of bulbar form that are troublesome in dealing with other people, such as saliva running from the mouth or uncontrolled crying, laughing or yawning.

End of life decisions

Some people diagnosed with ALS, feel the need for their wishes to be recorded early on, before the final stage of the disease, and take matters into their own hands. As the disease progresses, therapists usually urge that people decide about the respiratory policy. They want to prevent that someone suddenly is dependent on respiratory devices in a critical situation.
In practice, many people record their wishes in writing and already have spoken with their doctor about resuscitation, artificial respiration and often also about euthanasia. More than twenty percent of people with ALS dies by euthanasia.

Caregiving

Caregivers of people with ALS, especially the partners, frequently carry a heavy burden. A partner needs to get up during the night to help with going to the bathroom or with turning over in bed. The result of which may be a disturbed night rest and in the long term an overload. During the day, the help with personal hygiene also becomes more difficult when the muscles are no longer cooperating. The bulbar form of ALS creates significant obstacles for caregiving. The moment when homecare is called upon, the care often already has become complex and demanding. For that reason, it can become a disappointment for the caregivers if homecare does not sufficiently pay attention to the needs and emotions of the client and the care he or she needs. No matter the carefulness, the presence of strangers interrupts the privacy and rest at home. Some caregivers have the need for an aftercare by one of the home carers involved, after the

death of the person with ALS. For the caregiver, there is a high probability that he or she falls into a black hole. To process the intense years they have experienced, it appears to be important that they stay in contact with the home carers involved.

Motive to request home care
Rehabilitation teams or the Neuromuscular Reference Center advise patients to submit a request for home care on time. It occurs that they only do so if the care becomes too demanding or too technical for the caregiver at home. People want to spend the limited time they have together as meaningful, intense and dignified as possible. Leaning on professional help may lighten the burdens of the caregivers, but it is also a breach of privacy and intimacy. For that reason, a request for home care is sometimes put off as long as possible. At the moment, there are tests involving personal budget plans.

2. Acting responsible in a complex care situation

Points of concerns and practical advice

Dressing and undressing
– Because muscle function in people with ALS continues to deteriorate, the help required with dressing and undressing changes. Someone who could take off his or her own sweater the one week, may not anymore the other. Be alert to this.

- Someone who can no longer use his muscles to help, can no longer indicate whenever a movement hurts. Tell someone to indicate whenever a movement is uncomfortable.

- Listen to advice from family members or caregivers.

Treatment
Because of the speech difficulty and saliva loss, you may sometimes get a wrong impression of a person with ALS. Keep in mind that mental abilities of someone with ALS are not affected. Treat that person with respect and speak at a normal volume, their hearing is fine.

Eating, drinking and choking.
- When the muscles in the mouth and throat become weaker, problems may arise with eating and drinking. Chewing and swallowing will happen slower. It is important that the person with ALS receives enough nutrients. Take the time to help him or her with eating and drinking. Decide which position is most comfortable for him or her to eat or drink.

- When in doubt if the person with ALS is not eating or drinking enough, inform that person to contact a dietitian.

- Do not ask questions when a client is chewing or swallowing. Avoid saying funny or annoying things which may provoke a direct reaction.

- Due to the muscle weakness in the mouth, a person with ALS can choke easily. Pay attention with more fluid food and hard foods. The dietitian can give the advice to adapt the diet, for instance bread without the crusts, soft meats such as minced meat or the use of a thickening agent.

- The speech therapist of the specialist ALS treatment team can be called upon to provide tips on how to chew and swallow as safe as possible.

Coughing
- The reduced cough power can cause to an increase in the production of saliva or tightness in the chest and severe coughing fits. Stay with the person with ALS and remain calm. Encourage him or her to keep coughing until it is over.

- If there is a piece of food stuck in the oesophagus, use the Heimlich Manoeuvre.

- The physical therapist can give instructions about how to support the coughing.

Crying or laughing
Persons with ALS may sometimes start to laugh or cry with no reason. This compulsive crying or laughing is something they do not do on purpose. They experience this as annoying. The best thing to do is to indicate you are aware of that part of the disease and ask them in which way you can offer support. Medication can be prescribed if the compulsive crying or laughing becomes too troublesome, however, the medication may not be effective for everyone.

Cramps
- Ask the person with ALS what the most comfortable position is for him or her to avoid cramps.
- The physical therapist may provide instructions and tips to alleviate the cramping.
- Some patients receive medication from their rehabilitation specialist against the cramping, which may diminish the frequency of the cramps, but not the severity.

TIPS

Talking

- Take your time understanding someone who has difficulty talking. Make sure you know what he or she is saying and he or she knows what you are about to do.
- Make eye contact and be seated at eye level of the client. Repeat what you think it is you understand so the client does not have to repeat everything, only the word that is missing.
- Talk directly to the person with ALS.
- Try to mainly ask questions that do not require a long answer, such as yes and no questions.
- If someone is dependent on communication devices, make sure you are informed on how the device functions, so you can communicate through these devices with the patient. If that is too time consuming, talk to your superior.
- Questions about communication devices can be answered by a speech therapist.

Pain

- Pain can occur when someone stays in one position for too long, which causes the muscles and joints to stiffen. Make sure the person with ALS is comfortable and be aware of the pressure points.
- Pain can also occur because the muscles become overworked by weakening of other muscles. Consult a physio or occupational therapist to check if there are sufficient resources to avoid an overload.

Lifting and moving

- Since the muscle function of people with ALS keeps on deteriorating, the degree in which they can still assist in standing up and sitting down also changes. Be aware of that and adapt yourself to the help that is needed. Always keep the Individual care file up to date for the next caregiver.
- When the patient can no longer assist in lifting or moving and muscle weakness increases, they can literally slip through your hands. Assess the situation and make sure an extra pair of hands is around to help.
- Use resources that are present such as a lift or a sliding sheet. Make sure you know how to use them. When you need more time to do so, talk to your supervisor.
- In case of questions concerning lifting, moving or other resources, contact a physiotherapist or occupational therapist. Consult the contact list to see who is already involved with the client.

Falling

- A person with ALS has a greater risk of falling. If a fall leads to fractures and possible operations, this may worsen the disease. Encourage the person with ALS to use the available aids and home adaptations.
- Contact the care coordinator if you feel that the risk of falling is significant and adaptations need to requested.

Useful website: www.als.be

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