

# PALLIATIVE CARE OF PATIENTS WITH AMYOTROPHIC LATERAL SCLEROSIS

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## INTRODUCTION:

Amyotrophic lateral sclerosis (ALS) is a progressive disease of motoric nerve cells in the brain cortex (upper motoric neurons), and in the spinal cord and the medulla (lower motoric neurons). That is why ALS is sometimes also referred to as the Motor Neurone Disease (MND), although ALS actually is only one of the forms of MND. In other cases namely, either the lower or the upper motor neurones are damaged. In Europe, the eponymic name of the disease is sometimes used – Charcot's disease (after Jean Martin Charcot, who described the disease in the summer of 1869). In the USA the disease is referred to as Lou Gerhig's disease (after a famous baseball player who was stricken with MND in 1933).

Most commonly the muscular weakness, as a consequence of loss of motoric nerve cells is shown in the limbs (in 70 %), and more rarely (in 30 %) in swallowing and/or speech disorders. Men are more likely to contract the disease. The incidence of the disease grows with age, too. The disease's prevalence is 6 to 8 patients in a population of 100.000, and the incidence from 1,5 to 2 on 100.000 inhabitants annually. Five to 10% of the patients contract a hereditary form of the disease; in some 20% of these patients it is possible to establish a mutation on the gene for superoxide dismutation (SOD1). 2-3% of sporadic ALS patients also show the same genetic mutation.

The most typical first sign of the disease is the loss of strength in the muscles of one limb. The weakening then gradually affects the neighbouring muscles, then muscles on the other part of the body, and finally the muscles involved in breathing, speech and swallowing. Other signs of the disease include clumsiness in handling objects and a slowness of movements, fatigue, muscles atrophy, spasms, cramps, fasciculation in muscles and other. Signs showing that other parts of the brain cortex have been affected (e.g. problems with memorizing, intelligence, inner speech...) are exceptions. Eye movements, urinal and faecal continence and the sensory system are normally not affected. Problems that are not a direct consequence of muscles' weakness like depression and anxiety, sleeping disorders, constipation, hypersalivation, symptoms of chronic breathing weakness and pain can significantly decrease the quality of life in singular phases of the disease.

The origins of ALS are not known, and several hypotheses exist. Some of the possible explanations are excitotoxic damage of nerve cells, disorders of the immune system, infections, lack of neurotrophic factors, poisoning, disorders in hormonal activities and disorders in the activities of mitochondria. The only known cause is the already mentioned mutation on the *SOD1* gene. The excitotoxic theory is based on the hypothesis that the accumulation of excitatory neurotransmitters of glutamates causes the destruction of nerve cells. The immunological hypothesis assumes that

antibodies are created acting then against normal motoric nerve cells destroying them eventually. Viruses are also mentioned as the possible cause of ALS, among them the virus causing poliomyelitis, HIV and other. The poisons could include some chemical elements like lead, aluminium, manganese, calcium, selenium. Nerve cells need several neurotrophic factors for their development and existence; the lack of these factors could lead to a premature death of cells.

The ALS diagnosis is based on a clinical neurological examination. In order to confirm the diagnosis and in order to exclude other diseases, some other laboratory tests are run, most commonly electromyography, visual tests and tests of the cerebrospinal fluid. The diagnostic categories include convincing, probable, possible and suspect ALS.

The age of patients at the start of the disease is mostly over 40, the average age approximately 58 years. It is impossible to give precise predictions on the natural flow of the disease for each singular patient. Although ALS is considered a disease connected with short-time survival (an average of 3-4 years), it must be noted here that approximately 25% of the patients live for more than 5 years after the diagnosis, and 10% of the patients over 10 years.

There has been a real explosion of information on different views of the ALS in the last decade, ranging from pathogenesis to treatment. Patients' care has become more complex and some communities found special centres based on a multidisciplinary approach. So far, a drug that would stop or alter the course of the disease, does not exist. The drug Riluzol has been available since 1995, prolonging the average survival of ALS patients for several months. Despite of these facts, therapeutical nihilism is not justifiable. There are namely numerous ways of palliative cure that could significantly improve the quality of life of patients and their families. It is very important to stress out that palliative cure is not identical to symptomatic cures nor to the cure in the terminal phase of the disease. The palliative treatment is a multidisciplinary approach starting with communication the diagnosis and ending with family counselling during the time of dying and bereavement. Several experts are included in the process (work therapists, dieticians, physical therapists, speech therapists, nurses, psychologists, social workers, hospice employees, doctors and other volunteer lay persons). Many procedures in the palliative treatment have already been scientifically assessed. Cure recommendations have been drafted based on these analyses.

## DIAGNOSIS COMMUNICATION

This is the beginning of a palliative cure. A doctor, trusted by the patient, must communicate clearly in an open conversation that the disease may have different courses and that it is incurable;

the patient must be introduced to different forms of help. If the communication is ambiguous, the patient or his family might create their own false picture of the disease, often forcing the patient to search for different opinions and explanations from one physician to the other.

All necessary information today can also be found on web sites.

It is understandable that the physicians get confused in explaining that the disease is incurable. Despite the unease, this fact may not be left out or communicated only to the family (with exceptions in situations where it is known that the patient does not want to know diagnoses). The communication method always depends on the patient.

The communication starts only when the diagnosis has sufficient foundations. It is necessary to mention the name of the disease and explain it. Positive aspects of the disease must be mentioned, such as for instance that it basically does not inflict pain, that there are no disturbs of the perception, awareness or urination and that it is much researched disease. At the communication of the diagnosis and at any other decision on palliative treatment, the family members must be included. If they want a second opinion, they need help. They also need advice in case they want to search for help from a healer (alternative treatments).

When symptoms of dyspnoea appear or symptoms of chronic hypoventilations or when the vital capacity drops to under 50%, the patient should be informed that he is entering the final phase of the disease, that is the phase of dying. This is the right time for the discussion about artificial respiration, living will and the determination of a health care proxy.

### **MUSCLE WEAKNESS**

Muscle weakness is the main symptom of ALS. Active and passive exercise is used for the prevention of involuntary contractions and joint stiffness, but these activities may never cause exhaustion. As the disease progresses, the patients may use different orthopaedic aids (e.g. walking sticks, crutches, foot elevators, wheelchair) but also other aids increasing the patient's independence in everyday activities (such as special feeding aids, toilet seat aids, bath seat). It is necessary to introduce this topic early enough so that the patient and his family could get used to the idea of an aid and that they could accept the aid more easily when the time comes. Some patients have a short-term positive response regarding the increase of muscular strength by a treatment with acetylcholinesterase inhibitors.

### **FASCICULATION, MUSCULAR CONVULSIONS AND SPASMS**

Fasciculation in muscles are often the first symptom of the disease. They can develop into painful muscular convulsions. Spasticity, caused by the decay of upper motoric neurons, can sometimes be a major problem. Both can be successfully treated, the convulsions by quinidine (available as the drug SERECOR), by carbamazepine or phenytoin, and spasms by baclophen or tizanidine.

### **DYSARTHRIA**

The weakness of bulbar muscles causes, among other things, problems in articulation (dysarthria) and the patient's speech becomes hard to understand. Problems in communication are one of the severest consequences of the disease. Speech therapy is more

successful in patients with slow progression of the disease. Speech can be replaced by electronic gadgets, the use of alphabetical cards is also efficient.

### **DYSPHAGIA**

Due to problems in the movements of the tongue, the pharynx and the oesophagus the patient has problems with swallowing (dysphagia). The draughts cannot be controlled they can even reach the trachea (aspiration). Efficient help can be provided by adjusting the density of food and increasing its energy value (special collections of recipes are available), as well as learning different swallowing techniques. When this does not help and when the patient loses more than 10% of his body weight, it is necessary to reach a decision on the so called percutaneous endoscopic gastrostomy (PEG). This is a rather simple surgery performed under local anaesthesia, recommended before the vital capacity sinks under 50%.

### **DYSPNOEA**

Difficulties in breathing are the hardest symptom of ALS. When they occur, patients often react with anxiety. The first rule of palliative treatment of dyspnoea is to break the vicious circle dyspnoea-anxiety-dyspnoea. The presence of family members brings immediate comfort, the semi-seated position and respiratory physical therapy help, too. It is possible to decrease the subjective feeling of lack of air by morphine, and anxiety can be solved by lorazepam drugs for instance. If morphine is carefully dosed, it practically never depresses breathing.

As respiratory insufficiency progresses, symptoms of chronic night hypoventilation occur: fatigue and sleepiness at day, concentration problems, sleeping disorders, morning headaches, restlessness, tremor, increased perspiration, tachycardia, breathing with auxiliary breathing muscles, low appetite and weight loss, repeated inflammations of upper respiratory tracts, cyanosis, sight disorders, nausea, syncope and diffuse pain in the head, neck and limbs. These disorders can successfully be prevented by the usage of non-invasive intermittent (artificial) ventilation with a mask, which cannot always be applied in all patients. It is a temporary treatment aimed to an increase of the quality of life and not its prolongation. An alternative is the occasional administration of oxygen, which is worse choice due to the danger of depressing respiratory centres. A permanent solution can be artificial respiration by a tracheostomy, which is rarely the patients' choice.

### **COMPULSIVE LAUGHTER/CRYING**

This is a typical symptom of ALS, which must be distinguished from depression. It can be very embarrassing for patients if they are in company. The doctor should mention it and inform about treatment possibilities. The first choice drug is amitriptyline; dopamine and lithium can also be of help.

### **PSYCHOLOGICAL PROBLEMS**

The patients normally react by a reactory depression at the communication of the diagnosis. Psychotherapy can be of help in this period, ideally including the whole family. Although suicidal thoughts are rather common in patients suffering from ALS, suicides are rare. The most common cause of suicidal thoughts is the fear of becoming a burden to the family. The doctor should be

aware of depression symptoms in all phases of the disease, especially since they affect the survival and since there are efficient drugs against depression. Amitriptyline can, for instance, act as antidepressant and as a drug against sialorrhea, compulsive laughter/crying and sleeping disorders. It is necessary to follow the mental health of other family members, too.

### **SLEEPING DISORDERS**

The most common causes of sleeping disorders are anxiety, depression, inability to rotate in bed, fasciculation and muscular convulsions, dysphagia with aspiration and breathing disorders. The treatment should first take into consideration the cause(s) of the disorder. It is necessary to be careful in administering sedatives since they can depress breathing.

### **SALIVATION**

Although salivation is not the consequence of excessive generation of saliva, but of the weakness of facial muscles and the muscles involved in swallowing, this disorder is solved by drugs and procedures reducing the generation of saliva, e.g. by drugs (glycopyrrolate, amitriptyline, trichexifenidile, clonidine), by binding the salivary glands and by inactivation through irradiation or by botulin.

### **CONSTIPATION**

ALS is not a disease of the autonomous nervous system, and it does not affect the digestive tract. The constipation is usually the result of scarce moving and/or drugs administration (muscular relaxants, sedatives, anticholinergics, opiates). It can be cured by adjusting the diet, sufficient fluids and the use of laxatives.

### **PAIN**

ALS does not affect sensory nerve fibres transmitting pain, at least not clinically. The cause of muscular skeletal pain is the loss of muscular substance that used to »protect« bones and joints, and contractions of muscles and joints. Treatment consists in the use of non-steroid antirheumatics and physical therapy. Pain can also be caused by pressure on the skin due to a limited or absent ability to move. In this case it is very useful to often change the body's position. Should treatment with the named drugs fail, it is necessary to introduce opiates together with remedies against weakness (e.g. chlorpromazine) and laxatives.

### **PROBLEMS WITH MUCUS EXPECTORATION**

This is the symptom that is hardest to cure. Thicker mucus in the upper respiratory tracts is the consequence of decreased intake of liquids and lower ability of expectoration. The use of N-acetylcysteine rarely helps; the patient must drink more fluids, so that the mucus could become thinner, but in this way more mucus is produced and the problem is not solved. Mucus must usually be removed by suction or assisted expectoration. Physical treatment by vibrations is successful in the initial phases of the disease.

### **OTHER SYMPTOMS**

Due to the weakness of the diaphragm reflecting functionally on the lower oesophagus sphincter, the gastrooesophageal reflux develops. It is especially dangerous in PEG, when the reflux can also cause aspiration. It is cured by drugs for the strengthening of peri-

staltic and antacids. Since the weakened muscles no longer help in pumping vein blood, the limbs start swelling. Higher position of the legs, physical therapy and compression bandages help.

Bowel evacuation and frequent urination are signs of a spastic bladder. Oxibutinine helps.

Jaw pressuring can be a consequence of a pseudo bulbar injury; benzodiazepines help.

Laryngospasm, a sudden reflex squeezing vocal cords, can cause a sense of dyspnoea. Strong emotions, cold air, aspiration and other can be a trigger. It stops in a few seconds without intervention.

### **PSYCHOSOCIAL ASSISTANCE**

This is the most important aspect of palliative care, requiring a multidisciplinary approach. Patients' associations help, too. Psychosocial treatment of families is as important as the care for the patients. An especially sensitive problem are the needs and the fears of the patients' children and the assistance to patients in their role as parents.

### **SPIRITUAL CARE AND BEREAVEMENT**

The term spiritual has several meanings in this context and it is hard to define. This is the need of defining the person in his everyday life and can, but also need not, include religious contents. Such spirituality can help in accepting the disease and influence the decision regarding PEG and artificial respiration.

Spiritual help should also include the whole family, especially in decreasing problems during the time of bereavement. Research data show that the bereavement of family members in cases of ALS can be especially strong and long. It can begin as soon as at the moment of diagnosis communication (anticipatory bereavement).

### **TERMINAL PHASE OR THE PHASE OF DYING**

Studies show that the majority of ALS patients die peacefully, mostly in their sleep, there are no cases of suffocation. If the patients do not undergo artificial ventilation, the dying process starts with a gentle transition from sleep into a coma due to hypercapnia. It is necessary in this phase to make sure the dying person is not suffering: restlessness and signs of dyspnoea are cured by morphine; since morphine is not an anxiolytic, this drug should sometimes be added, too. The dosage should be sufficient enough to remove all the symptoms. The fear of respiratory depression is inappropriate in the dying phase.

The majority of patients express the wish of dying at home. The help of hospices is the most effective way of realizing that wish. Patients should contact the hospice early enough.

### **CONCLUSION**

ALS patients normally remain rational to the end of their lives. This can be scary for observers, and it can help hospitals in developing mechanisms of acceptance of the disease. The patients and their families usually want to actively participate in decisions on symptomatic treatments. It is the doctor's duty to establish such a partner relationship.

Science will probably find a cure that will stop the disease or at least prolong life. This will cause a higher prevalence of the disease, and consequently also an increase in the needs and demands for optimal palliative treatments.