Chapter 6

Model of care for a patient with amyotrophic lateral sclerosis (ALS) according to the International Classification for Nursing Practice (ICNP®)

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Abstract

Amyotrophic lateral sclerosis (ALS) is one of the most serious neurodegenerative diseases. It is a primary degenerative disease of the nervous system with a progressive course. ALS is a disease of unknown aetiology. For patients and families, the greatest cruelty of this disease is the fact that as it progresses, muscle atrophy and paresis occur. In the advanced stage, the sufferer is completely immobilised, unable to move despite full mental capacity and awareness. Although the disease is incurable, many of its symptoms can be alleviated, and the goal of therapy should be to improve the quality of life and maintain the patient's independence for as long as possible.

The aim of this study is to develop a model of care for a patient with ALS using the International Classification for Nursing Practice (ICNP®).

The study is based on an individual case study of a patient with ALS. Information for the development of the nursing model was collected on the basis of an interview with the patient and her family, observation of the patient, analysis

of medical documentation (patient's medical history, individual patient's order card), laboratory test results, assessment using scales (ALSFRS-R Scale, Barthel Index, Norton Scale), and measurements (blood pressure, pulse, temperature, and saturation). According to the model of individual nursing care, nursing diagnoses were made in accordance with the assumptions of the ICNP®, goals of care, planned nursing interventions and assessment of nursing care. This chapter aims to recognise the main health problems faced by the patient and her family. The nursing actions taken facilitated the care of the patient and improved the quality of her life that of her family.
Key words: amyotrophic lateral sclerosis (ALS), mechanical ventilation, model of care, ICNP®

Introduction

Amyotrophic lateral sclerosis (ALS) is an incurable progressive disease that leads to loss of muscle control. It belongs to the group of motor neuron diseases of unknown aetiology [1,2]. Its pathological process involves both the upper motor neurons (the motor cortex neurons) and peripheral motor neurons, including the alpha motor neurons of the ventral horns of the spinal cord as well as the nucleus of the brain stem. It results in progressive paresis, in various configurations, of the skeletal muscles, particularly the respiratory ones and those of the limbs, throat and larynx [1]. The onset of muscle weakness in ALS is typically focal and spreads to adjacent areas of the body. This is true of the spread of the disease within both the musculoskeletal system and segments of the spinal cord and the motor cortex. The disease typically presents through unilateral weakening of the distal muscles and atrophy of the muscles of the upper or lower limbs (spinal-onset ALS) or of the opponens pollicis. The disease gradually spreads to other anatomical regions. Its onset in the limbs is usually of an asymmetrical nature, affecting more often the upper limbs. It typically begins in the distal segment of the limb and causes atrophy of the small muscles of the hand. More rarely, atrophy affects the proximal muscles of the upper limbs [3]. ALS whose onset occurs in the opponens pollicis most often presents through dysarthria and dysphagia, and more rarely through dysphonia or reduced closure of the mouth or problems with swallowing. Weakening of the core muscles, head drooping and problems

with posture are frequent in later stages of the disease, but are rarely its main symptom. In some patients, weakening of the muscles is preceded by a period when bundle branch tremor, muscle cramps and slight weight loss are observed. ALS does not affect the senses and the intellect. It is also not associated with trophic disorders, dysfunction of the anal sphincter and bladder, nor with disorders of superficial and deep sensation. The rate of disease progression varies considerably from patient to patient. As a rule it is steady, but sometimes additional illnesses, such as a respiratory infection, can greatly speed it up. As a consequence, every ALS sufferer is able to understand and compose speech but is unable to speak. They completely lose the ability to move their limbs, they cannot swallow (aphagia) and cannot breath due to weakening of the respiratory muscles. They retain the ability to move their eyeballs and eyelids. Progressive respiratory failure together with increasing hypercapnia and coma is the most frequent cause of death in patients with ALS. In this situation, patients usually pass away peacefully, often in their sleep. Some patients die as a result of choking or aspiration pneumonia. The use of a respirator in order to support breathing can prolong survival, but it is not able to stop the progression of the disease [4].

A diagnosis of ALS is based on three principles [3,5]:

- Symptoms of functional impairment of a specific area of the body.
- The presence of signs of involvement of the central and peripheral motor neurons in one or more segmental anatomical areas.
- Progressive functional impairment.

Amyotrophic lateral sclerosis is diagnosed clinically and is supported by electrodiagnostic assessment. Electrodiagnostic tests are of key importance for a diagnosis of ALS because they detect the involvement of the lower motor neurons. Of particular importance is an electromyographic test (EMG) of the muscles as well as a conduction test in the motor and sensory fibres of the peripheral nerves (ENG) [3]. The EMG test and conduction test are basic diagnostic aids and serve both to identify diseases that mimic ALS and to reveal motor unit loss, which is the fundamental characteristic that defines the pathogenesis of the disease [3,6]. Each year between 1.5 and 2.5 people out of 100,000 develop ALS. From the moment

of the appearance of the first symptoms, the average life expectancy is two to four years, and the most common cause of death is respiratory failure. The dynamics of the progress of the disease varies greatly. This makes it impossible to predict the survival time or the point at which medical intervention will be indicated, the purpose of which is, first and foremost, to prevent the effects of dysphagia and failure of the respiratory muscles. In younger people, who are initially more mobile and have greater lung capacity, including when the first symptoms affect the muscles of the limbs, the disease progresses more slowly [1,6–8].

Case study

The patient is a 65-year-old retired clinical psychologist. She is married and the mother of three sons. She lives with her husband and one son on the third floor of a block of flats with a lift. In November 2020 she noticed that she was having progressive problems with speech. In June she consulted a neurologist for a diagnosis. The doctor interviewed her, performed some neurological tests and recommended an EMG test. The tests confirmed the diagnosis: ALS. From March 2021 the patient had experienced tongue fasciculations, weakening of the upper left limb, atrophy of the thenar muscles of the left thumb, paresis of the neck when flexing the muscles and fasciculations in the muscles of the upper limbs. In July 2021 she reported to the casualty department because of considerably increased dysphagia, she was experiencing choking, she was unable to take in fluids and, furthermore, she had lost 10 kg in six months. In the casualty department she was assessed by a neurologist, it was recommended that she be hospitalised, receive a surgical consultation and be prepared to have a PEG (percutaneous endoscopic gastrostomy) tube fitted. In December 2021 a tracheotomy procedure was performed on the patient due to shortness of breath and choking – mostly because of secretion blockage. Her husband, son and three carers are responsible for her day-to-day care. She is well-oriented in all aspects and her mood is stable. The patient has come to terms with her illness and she tries to enjoy every day. Her deep faith means that despite her illness she neither despairs nor loses hope. She has

a lot of support from her husband, sons, carers and friends. The patient is conscious, spends all her time in a lying position, and requires constant help from others in every aspect of her everyday functioning.

On the Barthel Index she was given 20 points.

On the Norton Scale she was given 10 points

On the ALSFRS-R Scale she was given 0 points.

She has been placed on a programme of invasive mechanical ventilation in a home setting. Ventilation is carried out twenty-four hours a day, invasively through a tracheostomy tube with an Astral respirator. The patient uses an adjustable electric rehabilitation bed with an anti-decubitus alternating pressure mattress. She expresses 'yes' by blinking her eyes and 'no' by nodding her head, and if she wants to communicate something with the aid of her carer she uses a chart with the letters of the alphabet or C-Eye. At the beginning of her care the patient was able to move her hand slightly, but despite the exercises performed by her physiotherapist and carers the disease quickly caused complete paralysis. She is lying in the Fowler position, suctioned if thick mucus accumulates, and has the bandage around her tracheostomy tube changed twice a day. The skin around the tracheostomy is clean, without the presence of granulation tissue and there is no discharge from the tube. The side on which she is lying is changed every two hours and during toileting. The patient reacts to voice and pain stimuli. She is aware of herself and her surroundings. She has a normal physique, she weighs 60 kg, she is 160 cm tall and her BMI is 23.44. She is fed a blended and commercial diet via the PEG and has full control over her sphincters, although she defecates into protective underwear. Apart from nursing care, the patient is under the care of an anaesthetist (who visits once a week) and a physiotherapist (twice a week). Care involves laboratory tests of the blood and urine. The family can summon the nurse by telephone twenty-four hours a day.

Nursing diagnoses

Diagnosis 1. Impaired function of the respiratory system [10023362], impaired gas exchange [10001177]

Client: patient
Date added: 25.06.2022
Aim of care: effective breathing [10041334], effective gas exchange [10027993]
Planned nursing interventions:
- monitoring the vital signs [10032113]
- monitoring respiratory therapy [10037092]
- monitoring respiratory function [10012196]
- teaching the family to monitor respiratory function [10036971]
- teaching to measure the respiratory rate [10044772]
- teaching to take the pulse on the wrist [10045550]
- teaching to measure the body temperature [10044738]
- suctioning of airways [10044890]
- oxygen therapy [10039369]
- encouraging the use of breathing or coughing techniques [10006834]
- using support for positioning [10035467]
- measuring respirations [10046338]
- maintaining airway patency [10037351]
- respiratory support device [10016958]: respirator [10044842]
- endotracheal tube [10006868]
- monitoring blood oxygen saturation using a pulse oximeter [10032047]
- sustaining respiration [10031674]
- ensuring constant care [10006966]

Evaluation of nursing care: properly functioning respiratory system [10028156], effective gas exchange [10027993]

Diagnosis 2. Risk of aspiration [10015024]
Client: patient
Date added: 25.06.2022
Aim of care: effective maintaining of airway patency [10027964]
Planned nursing interventions:
- respiratory support device [10016958], suction apparatus [10019029]
- manual aspiration [10011723]

– suctioning of airways [10044890]
– monitoring respiratory function [10012196]
– assessing respiratory status using a monitoring device [10002799]
– assessing behaviour relating to eating and drinking [10002747]
– positioning the patient [10014761]
– encouraging the use of breathing or coughing techniques [10006834]
– expectoration [10007362], sputum [10018717]

Evaluation of nursing care: no aspiration [10028783]

Diagnosis 3. Dysphasia [10006457]
Client: patient
Date added: 25.06.2022
Aim of care: ability to communicate effectively [10014790]
Planned nursing interventions:
– identification of barriers to communication [10009683]
– assessing verbal communication abilities [10030515]
– assessing the degree of the family's ability to cope [10026600]
– teaching about effective communication [10036169]
– encouraging effective communication in the family [10036066]
– facilitating the communication of needs [10038196]
– communication device [10004714], computer [10004906]
– instruction material [10010395], facilitating the communcation of needs [10038196]

Evaluation of nursing care: ability to communicate [10000052]

Diagnosis 4. Risk of pressure ulcers [10027337]
Client: patient
Date added: 25.06.2022
Aim of care: prevention of pressure ulcers [10029065]
Planned nursing interventions:
– therapy using a support device [10039158], anti-decubitus mattress [10041560]
– assessing the risk of pressure ulcers [10030710]

- teaching about care of pressure ulcers [10044218]
- teaching about preventing pressure ulcers [10036861]
- assessing pressure ulcers [10040847]
- preventing pressure ulcers [10040224]
- implementing care that ensures comfort [10039705]
- positioning the patient [10014761]
- moving the patient [10033188], lifting equipment [10011349]
- dietary management [10023861]

Evaluation of nursing care: no pressure ulcers [10029077]

Diagnosis 5. Impaired function of the musculoskeletal system [10022642]
Client: patient
Date added: 25.06.2022
Aim of nursing: provision [10015935], need [10012495]
Planned nursing interventions:

- therapy using a support device [10039158]
- providing safety equipement [10024527], a bed rail [1000320]
- evaluating the status of the musculoskeletal system [10034030]
- assessing mobility [10030527]
- positioning of the patient [10014761]
- assisting with mobility in bed [10045972]
- assisting with eating and drinking [10037269]
- assisting with self-care [10035763]
- assisting with hygiene [10030821]
- rehabilitation [10016645]
- techniques for exercising the muscles and joints [10012300]
- transcutaneous neurostimulation device [10019188]
- enhancing the techique for performing muscle and joint exercises [10036512].

Evaluation of nursing care: properly functioning musculoskeletal system [10012773]

Diagnosis 6. Self-care deficit [10023410]
Client: patient
Date added: 25.06.2022
Aim of care: ensuring continuity of care [10006966], the family's ability to cope [10034736]
Planned nursing interventions:
- assisting with toileting [10023531]
- assisting with hygiene [10030821]
- assisting with eating and drinking [10037269]
- assisting with mobility in bed [10045972]
- dressing the patient [10031164]
- implementing care that ensures comfort [10039705]
- teaching the family about hygiene practices [10038131]
- teaching about mouth care [10038108]
- teaching about care of the perineum [10045165]
- teaching about tracheostomy care [10044888]
- assessing self-care [10021844]
- assessing possibilities [10026040]
- assessing self-efficacy [10024280]
- assessing the degree of the family's ability to cope [10026600]
- assessing the carer [10030562]
- maintaining dignity and privacy [10011527]
- assessing attitudes towards the disease [10024192]

Evaluation of nursing care: effective continuity of care [10035507], ability of the family to cope effectively [10034770]

Diagnosis 7. Risk of infection [10015133] + respiratory tract [10002100]
Client: patient
Date added: 25.06.2022
Aim of care: prevent infection [10028945]
Planned nursing interventions:
- monitoring the objective and subjective symptoms of infection [10012203]

– monitoring blood oxygen saturation using a pulse oximeter [10032047]
– monitoring the temperature of the body [10012165]
– monitoring the blood pressure [10032052]
– teaching the family about susceptibility to infection [10038149]
– teaching the family about preventing infection [10036928]
– assessing the signs and symptoms of infection [10044182]
– assessing susceptibility to infection [10002821]
– preventing infection [10036916]
– using aseptic technique [10041784]

Evaluation of nursing care: no infection [10028945]

Diagnosis 8. Potentiality [10015151] + malnutrition [10042077]
Client: patient
Date added: 25.06.2022
Aim of care: improvement in nutritional status [10035569]
Planned nursing interventions:
– assessing the risk of impaired nutritional status [10040921]
– assessing the risk of dehydration [10040932]
– assessing the nutritional status [10030660]
– assessing adherence to recommendations [10024185]
– assessing attitudes towards nutritional status [10002694]
– monitoring nutrition [10036032]
– teaching the family about the dietary regimen [10026525]
– teaching about dietary requirements [10046533]
– teaching about dehydration [10043832]
– teaching about nutrition [10024618]
– teaching about enteral feeding [10046546]
– demonstrating how to administer medication [10024354]
– implementing enteral feeding [10046178]

Evaluation of nursing care: effective response to enteral nutrition [10033427], positive nutritional status [10025002]

Diagnosis 9. Constipation [10000567]
Client: patient
Date added: 25.06.2022
Aim of care: treatment of constipation [10044729]
Planned nursing intervention:
- assessing bowel function [10036475]
- assessing the risk of dehydration [10040932]
- assessing the nutritional status [10030660]
- teaching the family about the dietary regimen [10026525]
- teaching about dietary requirements [10046533]
- managing defecation [10041427]
- managing digestion [10031782]
- monitoring fluid intake [10035303]
- monitoring food intake [10036614]
- monitoring the response to treatment [10032109]

Evaluation of nursing care: effective defecation [10028403]

Diagnosis 10. The risk of the side effects of medication [10037604]
Client: patient
Date added: 08.07.2022
Aim of care: no side effects of medication [10040295]
Planned nursing interventions:
- administering medication [10025444]
- demonstrating how to administer medication [10024354]
- evaluating the response to medication [10007182]
- monitoring the side effects of medication [10043884]
- assessing adherence to recommendations [10024185]
- encouraging adherence to medication recommendations [10038051]
- teaching about techniques for reducing risk [10038804]
- teaching about the side effects of medication [10044614]
- teaching about medication [10019470]

Evaluation of nursing care: no side effects of medication [10040282]

Diagnosis 11. Depression [10022402]
Client: patient
Date added: 01.12.2022
Aim of care: improvement [10026692] + mood [10036241]
Planned nursing interventions:
- assessing depression [10026055]
- assessing the ability to cope [10002723]
- assessing the attitude towards the disease [10024192]
- providing emotional support [10027051]
- interaction with the family [10035887]
- conversation [10019436]
- motivation [10012242]

Evaluation of nursing care: reduced level of depression [10027901]

Summary

This case shows how essential nursing care is in treating a patient and ensuring an improvement in the quality life. It describes the most important health problems with which patients with ALS have to struggle at the various stages of their illness. It shows how important cooperation is between the nurse, doctor, physiotherapist and other people involved in the patient's care. A combination of appropriate communication, swift identification of troublesome symptoms and effective intervention create a chance for an improvement in the quality of life of patients with ALS. The nurse spends the most time with the patient during his/her stay in hospital as well as during his/her long-term care at home. This person plays an active role in pharmacotherapy, nursing, rehabilitation, preparing the patient for tests and taking samples for tests. S/he is the main support for the patient and the family and takes on a primary educational role. When the patient and the family experience difficulty while performing certain activities, for example suctioning or administering medication via the PEG, they know who they must turn to for help. The patient is under the constant care of an interdisciplinary team. In this case, all the interventions improved the quality of life of the patient and her family.

Conclusions

According to the model for individual nursing care, nursing diagnoses were made in line with the International Classification for Nursing Practice (ICNP®), the nursing aims were defined, nursing interventions were planned and a nursing care assessment was performed. The nursing procedures implemented facilitated care for the patient and improved the quality of her and her family's life. Accurate diagnoses were made, which suggested the direction that the nursing care should take, made targeted interventions possible and enabled the expected care outcomes to be defined. The patient and her family were provided with medical, psychological and educational support. The patient's family were informed about how to look after a person suffering from amyotrophic lateral sclerosis properly.

Bibliography

1. Butna K, Pyszora A, Adamczyk A, Krajnik M. *Practical aspects of nursing care provided to patients diagnosed with amyotrophic lateral sclerosis receiving home mechanical ventilation*. Palliative Medicine in Practice. 2021;15(1):42–52, https://doi.org/10.5603/PMPI.2021.0005.
2. Blacharska-Krzanowska KA, Strzępek KA, Woroń J, Weber-Lipiec T. *How to manage exacerbation of respiratory failure in a patient diagnosed with amyotrophic lateral sclerosis*. Palliative Medicine in Practice. 2020;14(1):44–50, https://doi.org/10.5603/pmpi.2020.0006.
3. Kozubski W, Liberski PP (red.). *Neurologia: podręcznik dla studentów medycyny*. Tom 2. Wyd. 2 rozsz. i uaktual., PZWL Wydawnictwo Lekarskie, Warszawa 2016.
4. Adamczyk A, Kwiatkowska M, Filipczak-Bryniarska I. *Stanowisko Polskiego Towarzystwa Medycyny Paliatywnej dotyczące kwalifikacji do opieki paliatywnej i postępowania z pacjentem ze stwardnieniem zanikowym bocznym i stwardnieniem rozsianym*. Medycyna Paliatywna. 2018;10(3):115–130, https://doi.org/10.5114/PM.2018.79834.
5. Kozubski W, Liberski PP (red.). *Neurologia: podręcznik dla studentów medycyny*. Tom 1. Wyd. 2 rozsz. i uaktual., PZWL Wydawnictwo Lekarskie, Warszawa 2016.
6. Puchała Ł, Maksymowicz S. *Kierunki poszukiwań nowych leków w SLA*. Farmacja Polska. 2019;75(5):247–250, https://doi.org/10.32383/farmpol/116234.

7. Rubinowicz-Zasada M, Orczyk A, Orczyk M, Pasek J. *Stwardnienie boczne zanikowe – choroba neuronu ruchowego. Prezentacja przypadku*. Pediatria i Medycyna Rodzinna. 2015;11(1):112–118, https://doi.org/10.15557/PIMR.2015.0010.
8. Louis ED, Mayer SA, Rowland LP (red.). *Merritt Neurologia*. Tom 2. Red. wyd. pol. i tłum. W Turaj. Wyd. 4, Edra Urban & Partner, Wrocław 2018.