An educational programme for home mechanical ventilation in motor neuron disease

Federica Riva-Rovedda[1], Marco Clari[1], Alessio Conti[2], Tiziana Esposito[3], Giuseppe Tabbia[4], Alessio Mattei[5], Andrea Calvo[6], Paolo Solidoro[4,7] and Valerio Dimonte[1]

[1]Department of Public Health and Pediatrics, University of Torino, Turin, Italy. [2]Department of Clinical and Biological Sciences, University of Torino, Turin, Italy. [3]Pneumology U, Respiratory Rehabilitation Service, Città della Salute e della Scienza University Hospital, Turin, Italy. [4]Pneumology U, Città della Salute e della Scienza University Hospital, Turin, Italy. [5]Pneumology, Santa Croce e Carle Hospital, Cuneo, Italy. [6]ALS Center, Department of Neuroscience "Rita Levi Montalcini", University of Torino, Turin, Italy. [7]Department of Medical Sciences, University of Turin, Turin, Italy.

Corresponding author: Marco Clari (marco.clari@unito.it)

Shareable abstract (@ERSpublications)
Managing home mechanical ventilation is a challenge for people with MND and their informal caregivers. The educational programme proposed by the Regional Expert Centre of a Northern Italy university hospital could facilitate everyday care. https://bit.ly/4gXSJML

Cite this article as: Riva-Rovedda F, Clari M, Conti A, *et al*. An educational programme for home mechanical ventilation in motor neuron disease. *Breathe* 2025; 21: 240094 [DOI: 10.1183/20734735.0094-2024].

Received: 14 May 2024
Accepted: 1 Oct 2024

Abstract

Respiratory failure represents the leading cause of death among people with motor neuron disease (MND). Home mechanical ventilation supports respiratory function, but its management places a significant demand on patients and informal caregivers. The need for consistent support to deal with home mechanical ventilation is recognised. Regardless, specific educational interventions are infrequently reported. Within its multidisciplinary clinical care pathway, the Regional Expert Centre for MND of a Northern Italy university hospital offers patients and their caregivers an educational programme on how to deal with noninvasive ventilation (NIV) and invasive mechanical ventilation (IMV) at home.

NIV education is usually performed in an outpatient setting. Pneumologists and nurses train patients and caregivers about ventilator functions, management of respiratory interfaces, inhalation therapy, skin integrity preservation and respiratory deterioration detection, while respiratory therapists train patients and caregivers about the use of an assisted cough device. IMV education is performed at the hospital. Respiratory therapists, supported by nurses, train caregivers about ventilator functions, management of tracheostomy cannula, tracheal suction, inhalation therapy and use of assisted cough devices. Finally, a certification of the skills acquired is released. Future studies should assess the effectiveness of this programme in terms of satisfaction and clinical outcomes.

Background

Motor neuron disease (MND) is a rare and incurable disease characterised by progressive degeneration of both the upper and lower motor neurons [1]. Average survival does not usually exceed 3–5 years after the onset of symptoms [2]. People with MND experience a reduction in functional ability, lack of coordination of movements, spasticity, weakness and progressive muscular atrophy [1]. Consequently, they suffer from neurological, musculoskeletal, respiratory, nutritional and psychosocial impairments [1]. Among these, progressive respiratory failure is one of the most severe, representing the leading cause of death in MND [1].

Mechanical ventilation (MV) represents a key intervention to support respiratory function [3]. It can be administered as noninvasive ventilation (NIV) or invasive mechanical ventilation (IMV), and the two methods are provided to up to 44% and 31%, respectively, of people with MND in Europe [4]. Initially, NIV can prevent nocturnal hypoventilation, relieve respiratory muscles, and ensure optimal oxygen and carbon dioxide levels [5]. The progression of MND requires an increase in NIV sessions and, in the worst cases, a switch to IMV by tracheostomy. Despite MV, respiratory failure worsens over time, becoming fatal [3].

From diagnosis and throughout the progression of the disease, informal caregivers play an essential role in responding to the complex needs of people with MND in the community [6], including the management of MV when respiratory support is needed [7].

Multidisciplinary clinical care pathways are recommended to improve survival and quality of life while reducing hospital readmissions [8–10]. These pathways optimise access to care and cover the wide range of needs of people with MND and their caregivers [5]. Education and training in technical skills, such as MV, are often unmet needs for people with MND and their caregivers [11, 12]. However, educational interventions are infrequently reported [13, 14]. To date, most studies have focused on the acceptance, adaptation, characteristics and impact of MV on quality of life in people with MND and their caregivers [15–17]. Thus, this article aims to describe the clinical and educational programme for respiratory function follow-up and management of home mechanical ventilation (HMV) of a multidisciplinary clinical care pathway directed at people with MND and their informal caregivers.

Context

Since 2009, the Regional Expert Centre for MND (CRESLA) of the A.O.U Città della Salute e della Scienza University Hospital of Turin (Italy) has focused its activities on the epidemiological, clinical, cognitive, genetic, neuropathological and neurobiological aspects of MND. In 2022, CRESLA defined the multidisciplinary clinical care pathway for the management of MND, including the educational programme for the management of HMV developed by the Pneumology and Respiratory Rehabilitation Services of the hospital from 2018.

Clinical care pathway

Diagnosis

As no specific test has been identified to date, diagnosis is carried out by exclusion on the basis of signs and symptoms of motor neurons degeneration (*e.g.* muscle weakness, fasciculations), neurophysiological and neuroimaging investigations (*e.g.* electromyography, magnetic resonance imaging, positron emission tomography) and laboratory tests (*e.g.* C-reactive protein, liver function, protein electrophoresis). Diagnosis can be performed during neurology outpatient visits or in hospital when MND has an abrupt onset.

When patients receive the diagnosis, available treatments, necessary assessments and follow-up are discussed. At this stage, genetic counselling is planned to identify specific genetic mutations susceptible to cutting-edge targeted therapies [18]. When the form of MND developed by a patient has a genetic basis, the odds of MND recurrence among family members are defined and a transmission risk calculation is performed. Together with genetic counselling, a neuropsychological assessment is performed as >50% of people with MND experience cognitive-behavioural impairments and up to 15% are diagnosed with frontotemporal dementia [19].

After diagnosis, the neurology outpatient clinic is responsible for follow-up and shared care planning. Patients can access multiple clinical consultations, from pulmonologists, phoniatricians and physiatrists, to detect impairments early on. Focusing on respiratory care, people with MND follow a specific pathway to access the hospital outpatient clinic for home ventilation (figure 1).

Outpatient clinic for home ventilation

Respiratory function is assessed regularly by pulmonologists, nurses and respiratory therapists through spirometry, arterial blood gas analysis, polysomnography, evaluation of signs and symptoms, and nocturnal capnography. The results of these assessments are compared to well-defined criteria [5], in order to promptly highlight the need for MV support (table 1). Moreover, when the patient's cough results are ineffective, airway protection is compromised. Consequently, an assisted cough device is prescribed. When respiratory function assessments suggest NIV should be initiated, adaptation is proposed and accomplished over 2–3 pulmonologist visits over a short period of time.

Education

NIV

Since 2018, approximately 45 patients and their caregivers per year have been trained to deal with NIV at home. Education and training are carried out during the two visits where adaptation to NIV is performed for a mean duration of 45–60 min on each visit in the outpatient setting. A third appointment is scheduled if needed and during every follow-up visit education is consolidated. Pulmonologists and nurses describe the functioning, maintenance and management of the ventilator. Patients and caregivers are trained regarding the characteristics of the ventilator, ventilation timing and method of circuit assembly and replacement, with active and/or passive humidifiers. In addition, nurses help patients to choose the most

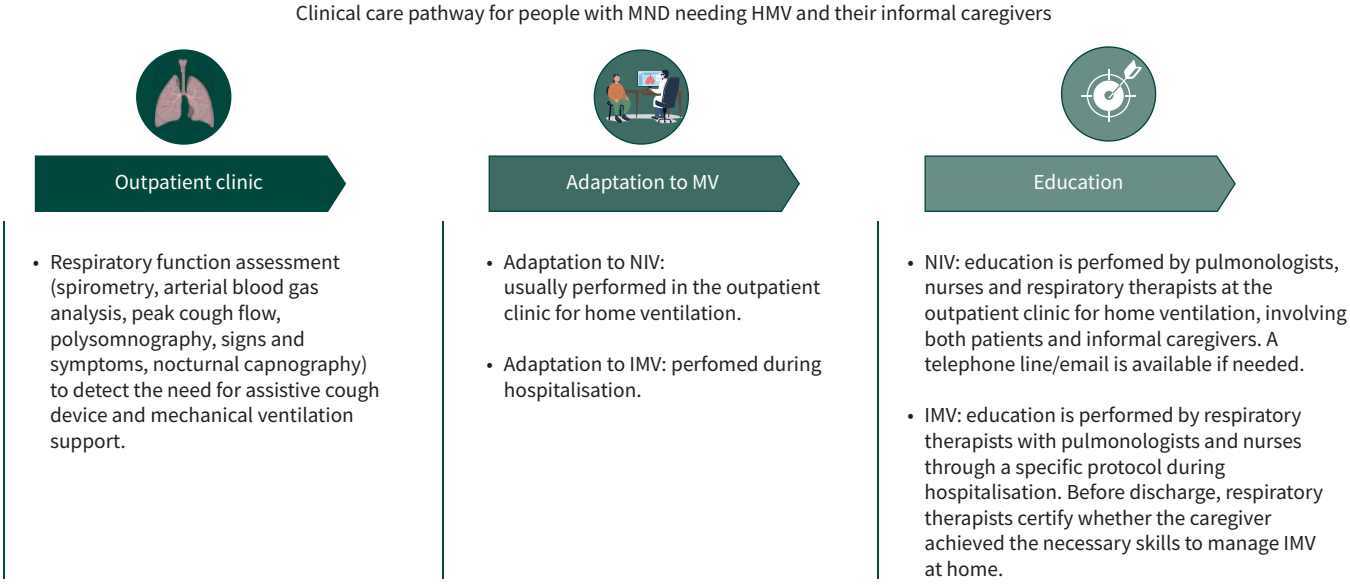

FIGURE 1 Clinical care pathway for people with motor neuron disease (MND) needing home mechanical ventilation (HMV), and their informal caregivers. MV: mechanical ventilation; NIV: noninvasive ventilation; IMV: invasive mechanical ventilation.

suitable mask by evaluating which mask is best tolerated. Furthermore, they describe how to preserve skin integrity, recognise and manage air leaks and how to cope with airway obstruction with support from respiratory therapists. They also train patients and informal caregivers about the correct positioning of the respiratory interface, connection to the oxygen, administration of inhalation therapy, airway clearance, and early detection of respiratory infections through the evaluation of signs and symptoms (*e.g.* increased body temperature, worsening dyspnoea). When an assistive cough device is required, respiratory therapists train patients on how to use the device effectively.

TABLE 1 Criteria for initiation of mechanical ventilation

Criteria for NIV initiation
Abnormal respiratory function tests; at least one of the following:
Forced vital capacity <80% of predicted value
Sniff nasal pressure <40 cmH_2O
Maximal inspiratory pressure <60 cmH_2O
Significant nocturnal desaturation
Morning blood gas P_{CO_2} >45 mmHg
Symptoms and/or signs related to respiratory muscle weakness; at least one of the following:
Dyspnoea
Tachypnoea
Orthopnoea
Disturbed sleep due to nocturnal desaturation
Morning headache
Use of auxiliary respiratory muscles
Paradoxical respiration
Daytime fatigue
Excessive daytime sleepiness
Criteria for assisted cough device prescription
Peak expiratory cough <270 $L \cdot min^{-1}$
Criteria for IMV initiation
Increased bronchial secretions that cannot be managed with noninvasive care methods
Increased dependency on NIV (>20 $h \cdot day^{-1}$)
Failure of NIV in providing effective ventilator pressure and gas exchange
Presence of severe swallowing impairment leading to chronic aspiration and recurrent episodes of pneumonia

P_{CO_2}: carbon dioxide tension; NIV: noninvasive ventilation; IMV: invasive mechanical ventilation.

Follow-up is scheduled every ∼3 months to monitor respiratory function not only on the basis of respiratory assessment tests, but also by analysing the data recorded by the patient's ventilator. Depending on the respiratory compensation, the timing of follow-up can vary, but a telephone line and an e-mail address are available in case of an emergency or doubts.

Education was never reported to be unsuccessful. Since 2018, 5–10% of patients chose to withdraw NIV because they did not tolerate it and did not report burdening respiratory symptoms. Nevertheless, they still felt confident to manage ventilation independently.

IMV

The progression of MND is associated with an increased dependence on MV, with the need to switch from NIV to IMV through a tracheostomy if the patient is willing to accept it. The presence of advance care planning expressed by the patient is pivotal because if the patient decides to accept IMV, a hospital admission is scheduled, and an otolaryngologist performs a tracheostomy before a sudden episode of respiratory failure arises. In the absence of a specific expressed wish, a tracheostomy is performed in the case of an emergency.

Afterwards, pulmonologists select the most suitable tracheostomy cannula, ventilator, ventilation parameters, type of humidification and ventilation circuit needed. During hospitalisation, nurses train caregivers about tracheostomy management, and respiratory therapists provide a structured educational programme to manage IMV only when patients prefer to be discharged at home and there is at least one caregiver willing to learn how to deal with IMV (table 2). Since 2018, approximately 10 patients per year have undergone a tracheostomy. For each patient, on average, two informal caregivers (family members and/or hired non-professional caregivers) are usually trained for 1 h per day over a mean of 3 weeks of hospitalisation. The educational programme is mainly directed to informal caregivers, but patients are always involved according to their residual functional abilities. If the caregiver does not feel confident in managing the tracheostomy independently, the patient cannot be discharged.

In addition, patients and caregivers are provided with a leaflet summarising the key messages of the education provided (*e.g.* synthesis of the characteristics of the tracheostomy cannula, aim and aspects of MV, humidification and oxygen therapy). The leaflet has a dedicated section on monitoring the clinical stability of patients, providing education on how to detect signs and symptoms of respiratory tract infections (*e.g.* observe whether the patient is using their neck muscles to breathe; check the aspect of tracheal secretions; read the oximetry value; check the values of respiratory rate, peak pressure and tidal volume; look for leaks on the ventilator). Another section is dedicated to the daily management of the ventilator, for example, reading and solving the alarms reported by the ventilator, with a list of the most frequently asked questions (*e.g.* Can the patient take a shower? Do I have to stimulate the patient to breathe on their own? Can I secure the catheter mount to the tracheostomy cannula at night to avoid accidental disconnection from the ventilator?) The leaflet allows healthcare professionals to personalise recommendations by filling dedicated blanks (*e.g.* number of assisted cough device sessions, peak pressure value) Lastly, respiratory therapists certify whether the patient and/or caregivers have achieved the skills to manage HMV independently prior to discharge.

The hospital outpatient clinic schedules the replacement of the tracheostomy cannula every ∼40 days. Informal caregivers are trained to perform this procedure only in the case of a genuine emergency (*e.g.* tracheostomy cannula displacement). The appointment for the first cannula replacement is also an opportunity to consolidate education.

Since education is performed only when informal caregivers highly motivated to care for the patients at home are present, to date, IMV education was never found to be unsuccessful. However, if informal caregivers report feeling overwhelmed due to the management of ventilation (both for NIV and IMV) at home or if at the end of the education they realise they are not able to provide a sufficient level of care, healthcare professionals help them to activate the support of hired non-professional caregivers or to evaluate the possibility of moving the patient to a specialised long-term care facility.

Discussion

This article presented the educational programme for the management of HMV practised by the multidisciplinary clinical care pathway of CRESLA. While the literature emphasises the importance of early referral for MV, there is a lack of uniformity on the optimal timing to start ventilation support as the timeline of MND progression is variable. CRESLA healthcare professionals discuss the MV options in advance with a personalised approach based on the inclusion of people with MND and their families to

TABLE 2 Educational content and skills acquired by informal caregivers through the educational programme to manage invasive mechanical ventilation at home

General information
Discharge programme
Prescribed aids (assistive devices)
Identification of healthcare professionals to contact in case of need or emergency
Management of clinical changes
Tracheostomy cannula
Description of the characteristics of the tracheostomy cannula
Inflating and deflating the tracheostomy cuff
Timing of inflation and deflation of the tracheostomy cuff
Description of the characteristics of the inner cannula
Emergencies
Tracheal cannula removal and replacement
Tracheal suction
Purpose
Characteristics of suction tubes
Checking of suction need
Suction technique
Regulation of suction pressure
Checking suction effectiveness
Equipment disposal
Suction system
Characteristics of the suction system
Correct suction pressure
Assembling the suction system
Correct maintenance of the suction system
Mechanical ventilator
Delivery and explanation of the manual for the ventilator
Timing of ventilation administration (diurnal, nocturnal, continuous, as needed)
Powering on and powering off the ventilator
Managing the power supply of the ventilator
Ordinary maintenance of the ventilator (*e.g.* dust filter replacement, cleaning)
Assembling monotube circuit with expiratory valve
Assembling ventilator circuit with active humidifier
Assembling ventilator circuit with passive humidifier
Management of ventilator equipment
Patient connection to the ventilator
Recognising and managing the alarms on the ventilator
Checking of ventilation mode
Checking of ventilation parameters
Checking of set up alarms
Alternating the ventilation mode
Rotation and activation of the back-up ventilator
Managing malfunctions of the mechanical ventilator
Handling power outage emergencies
Ventilator–patient circuit
Characteristics, timing and technique of replacement of the catheter mount
Characteristics, timing, technique of replacement, and assembling with active humidifier of the two-way circuit
Characteristics, timing, technique of replacement, and assembling with passive humidifier of monotube circuit with expiratory valve
Patient connection to the ventilator
Home clinical monitoring
Oximetry
Respiratory rate, tidal volume and peak pressure
Humidification
Assembly, use and maintenance of the active humidifier
Assembly, use and replacement of the passive humidifier

Continued

TABLE 2 Continued

Inhalation therapy
Inhalation therapy administration techniques
Checking of inhalation therapy device knowledge and use
Proper placement in the ventilator circuit of inhalation therapy
Use of a spacer for inhalation therapy
Oxygen therapy
Use of specific equipment for oxygen delivery (*e.g.* reservoir)
Connecting oxygen using the auxiliary manual breathing unit
Connecting oxygen using the ventilator
Airway clearance
Airway clearance technique performance
Detection of respiratory tract infections
Increased body temperature
Ventilator alarms
Increased number of tracheal suctions
Change in secretions appearance

ascertain the best time for MV according to the patient's general condition and not in relation to respiratory function parameters alone.

The CRESLA clinical care pathway is a genuine example of a multidisciplinary approach, as it does not merely list a series of medical assessments but sets out structured multiprofessional integrated evaluations and follow-up interventions. In fact, respiratory function is evaluated not only by pulmonologists and respiratory therapists but also by phoniatrists and speech therapists to detect the presence of potential risk factors that could deteriorate respiratory function. This structured approach allows regular monitoring of respiratory function, which is known to promptly identify the risk of patient decompensation and provide an opportunity for early referral to advance care planning [20]. Consequently, if the patient is willing to accept IMV, a tracheostomy could be scheduled to avoid emergency procedures, which expose patients to negative clinical outcomes.

Formal education allows caregivers to provide care at home effectively and safely [7]. In fact, to date, there is no clear evidence that home care provided by professional caregivers is superior to informal care [21]. From our results, all the caregivers that decided to be educated succeed in caring for their relatives at home. Education increased their awareness of their relatives' needs and recognition of the importance of asking for help when feeling overwhelmed.

Education is essential to discharge patients with medical needs at home, but it should be specified that preferring home as a place of care is never a choice dictated by an inferior socioeconomic status. As a universal public system, the Italian healthcare system does not charge patients and the welfare system of the country provides financial support to afford long-term specialist care or to hire a non-professional caregiver when being discharged at home is not possible or a family caregiver is not present.

The presence of a certificate released by healthcare professionals within the educational programme represents a particular strength, as it validates the skills acquired, improving personal confidence and fostering empowerment. Besides, formal education also has legal and ethical implications to consider. First, formal education safeguards the free choice of people with MND and their informal caregivers to be discharged home, not forcing patients to move in to a residential facility, in agreement with the Charter of fundamental rights of the European Union [22], the Italian Constitution [23] and Italian Law n.219/2017 [24] concerning the right of free choice and self-determination. Secondly, the provision of formal and structured education relieves healthcare professionals from any responsibility in case of patients being harmed because of informal caregivers assistance. Lastly, considering the will of the patients and the ethical principles of beneficence (doing good), non-maleficence (to do no harm), autonomy (giving the patient the freedom to choose freely) and justice (ensuring fairness), formal education is the pillar that allows supporting managing care at home.

Caring for a person with MND represents an overwhelming experience, especially because often caregivers report being poorly trained for their role, contributing to an increase of their burden [25]. Consequently, improving the quality of care through specific educational programmes is crucial both for the quality of life of the patients and their informal caregivers.

Conclusion

The educational programme of the CRESLA clinical care pathway represents an actual effort to support people with MND undergoing HMV and their informal caregivers, facilitating everyday home care. Future research should assess the efficacy of education and training in terms of clinical outcomes, satisfaction and quality of life.

Conflict of interest: None of the authors has any potential financial conflict of interest related to this manuscript. Outside the submitted work, M. Clari reports being on the editorial board for *Breathe* (unpaid).

References

1 Ng L, Khan F, Young CA, *et al.* Symptomatic treatments for amyotrophic lateral sclerosis/motor neuron disease. *Cochrane Database Syst Rev* 2017; 1: CD011776.
2 Morgan S, Orrell RW. Pathogenesis of amyotrophic lateral sclerosis. *Br Med Bull* 2016; 119: 87–98.
3 Radunovic A, Annane D, Rafiq MK, *et al.* Mechanical ventilation for amyotrophic lateral sclerosis/motor neuron disease. *Cochrane Database Syst Rev* 2017; 10: CD004427.
4 Turner MR, Faull C, McDermott CJ, *et al.* Tracheostomy in motor neurone disease. *Pract Neurol* 2019; 19: 467–475.
5 EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis: Andersen PM, Abrahams S, Borasio GD, *et al.* EFNS guidelines on the clinical management of amyotrophic lateral sclerosis (MALS)—revised report of an EFNS task force. *Eur J Neurol* 2012; 19: 360–375.
6 de Wit J, Bakker LA, van Groenestijn AC, *et al.* Caregiver burden in amyotrophic lateral sclerosis: a systematic review. *Palliat Med* 2018; 32: 231–245.
7 Bassola B, Cilluffo S, Lusignani M. Going inside the relationship between caregiver and care-receiver with amyotrophic lateral sclerosis in Italy, a Grounded Theory study. *Health Soc Care Community* 2021; 29: 1083–1090.
8 Hogden A, Foley G, Henderson RD, *et al.* Amyotrophic lateral sclerosis: improving care with a multidisciplinary approach. *J Multidiscip Healthc* 2017; 10: 205–215.
9 Martin S, Trevor-Jones E, Khan S, *et al.* The benefit of evolving multidisciplinary care in ALS: a diagnostic cohort survival comparison. *Amyotroph Lateral Scler Frontotemporal Degener* 2017; 18: 569–575.
10 Berlowitz DJ, Mathers S, Hutchinson K, *et al.* The complexity of multidisciplinary respiratory care in amyotrophic lateral sclerosis. *Breathe (Sheff)* 2023; 19: 220269.
11 Oh J, Kim J. Information-seeking behavior and information needs inpatients with amyotrophic lateral sclerosis: analyzing an online patient community. *Comput Inform Nurs* 2017; 35: 345–351.
12 Chu HS, Oh J. Information needs and preferences of family caregivers of patients with amyotrophic lateral sclerosis. *J Neurosci Nurs* 2020; 52: 207–213.
13 Kavanaugh MS, Howard M, Banker-Horner L. Feasibility of a multidisciplinary caregiving training protocol for young caregivers in families with ALS. *Soc Work Health Care* 2018; 57: 1–12.
14 Balañá A, Rubio MÁ, Bertran B, *et al.* Telematic adaptation to home mechanical ventilation in patients with amyotrophic lateral sclerosis. *Neurologia (Engl Ed)* 2023; 38: 59–61.
15 Winther D, Kirkegaard Lorenzen C, Dreyer P. Everyday life experiences of close relatives of people with amyotrophic lateral sclerosis receiving home mechanical ventilation–a qualitative study. *J Clin Nurs* 2020; 29: 2306–2316.
16 Kim CH, Kim MS. Ventilator use, respiratory problems, and caregiver well-being in Korean patients with amyotrophic lateral sclerosis receiving home-based care. *J Neurosci Nurs* 2014; 46: E25–E32.
17 Pagnini F, Banfi P, Lunetta C, *et al.* Respiratory function of people with amyotrophic lateral sclerosis and caregiver distress level: a correlational study. *Biopsychosoc Med* 2012; 6: 14.
18 Grassano M, Calvo A, Moglia C, *et al.* Systematic evaluation of genetic mutations in ALS: a population-based study. *J Neurol Neurosurg Psychiatry* 2022; 93: 1190–1193.
19 Rusina R, Vandenberghe R, Bruffaerts R. Cognitive and behavioral manifestations in ALS: beyond motor system involvement. *Diagnostics (Basel)* 2021; 11: 624.
20 Baxter SK, Johnson M, Clowes M, *et al.* Optimizing the noninvasive ventilation pathway for patients with amyotrophic lateral sclerosis/motor neuron disease: a systematic review. *Amyotroph Lateral Scler Frontotemporal Degener* 2019; 20: 461–472.
21 Coe NB, Konetzka RT, Berkowitz M, *et al.* The effects of home care provider mix on the care recipient: an international, systematic review of articles from 2000 to 2020. *Annu Rev Public Health* 2021; 42: 483–503.
22 European Parliament, European Council, European Commission. Charter of fundamental rights of the European Union. Official Journal of the European Union. Date last accessed: 1 August 2024. Date last updated: 2016. https://eur-lex.europa.eu/eli/treaty/char_2016/oj
23 Presidenza del consiglio dei ministri. Italian Republic Constitution. Date last accessed: 1 August 2024. Date last updated: 7 October 2023. www.normattiva.it/uri-res/N2Ls?urn:nir:stato:costituzione:1947-12-27!vig

24 Law of the Italian Republic 22 december 2017, n. 219 Norme in materia di consenso informato e di disposizioni anticipate di trattamento. [Regulations on informed consent and advance treatment arrangements]. Date last accessed: 3 August 2024. Date last updated: 31 January 2018. www.gazzettaufficiale.it/eli/id/2018/1/16/18G00006/sg

25 Law S, Ormel I, Babinski S, *et al.* "Caregiving is like on the job training but nobody has the manual": Canadian caregivers' perceptions of their roles within the healthcare system. *BMC Geriatr* 2021; 21: 404.