

Taylor & Francis
Taylor & Francis Group

Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration

ISSN: 2167-8421 (Print) 2167-9223 (Online) Journal homepage: www.tandfonline.com/journals/iafd20

End-of-life care of patients with amyotrophic lateral sclerosis and home mechanical ventilation: the national population-based DISCOVERY study

Mikaela Dalkvist , Karin Ersson, Stefan Bergström , Magnus Ekström, Össur Ingi Emilsson , Ludger Grote, Caroline Ingre, Ulf Kläppe, Staffan Lundström, Ingela Nygren, Josefin Sundh & Andreas Palm

To cite this article: Mikaela Dalkvist , Karin Ersson, Stefan Bergström , Magnus Ekström, Össur Ingi Emilsson , Ludger Grote, Caroline Ingre, Ulf Kläppe, Staffan Lundström, Ingela Nygren, Josefin Sundh & Andreas Palm (21 Oct 2026): End-of-life care of patients with amyotrophic lateral sclerosis and home mechanical ventilation: the national population-based DISCOVERY study, Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration, DOI: 10.1080/21678421.2026.2747563

To link to this article: https://doi.org/10.1080/21678421.2026.2747563

© 2026 The Author(s). Published by Informa UK Limited, trading as Taylor & Francis Group.

View supplementary material

Published online: 21 Oct 2026.

Submit your article to this journal

Article views: 336

View related articles

View Crossmark data

Full Terms & Conditions of access and use can be found at
https://www.tandfonline.com/action/journalInformation?journalCode=iafd20

Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration, 2026; 0: 1–8

Taylor & Francis
by informa

OPEN ACCESS Check for updates

RESEARCH ARTICLE

End-of-life care of patients with amyotrophic lateral sclerosis and home mechanical ventilation: the national population-based DISCOVERY study

MIKAELA DALKVIST[1#], KARIN ERSSON[1,2,3#], STEFAN BERGSTRÖM[4], MAGNUS EKSTRÖM[5], ÖSSUR INGI EMILSSON[1,6,7], LUDGER GROTE[8,9], CAROLINE INGRE[10,11], ULF KLÄPPE[10,11], STAFFAN LUNDSTRÖM[12,13], INGELA NYGREN[14], JOSEFIN SUNDH[15] & ANDREAS PALM[1]

[1]Department of Medical Sciences, Respiratory, Allergy and Sleep Research, Uppsala University, Uppsala, Sweden, [2]Department of Medical Sciences, Clinical Physiology, Uppsala University, Uppsala, Sweden, [3]Department of Women's and Children's Health, Physiotherapy, Uppsala University, Uppsala, Sweden, [4]Department of Immunology, Genetics and Pathology, Uppsala University, Uppsala, Sweden, [5]Department of Clinical Sciences, Respiratory Medicine, Allergology and Palliative Medicine, Faculty of Medicine, Lund University, Sweden, [6]Faculty of Medicine, University of Iceland, Reykjavik, Iceland, [7]Department of Respiratory Medicine, Landspitali, Reykjavik, Iceland, [8]Centre for Sleep and Wake Disorders, Sahlgrenska Academy, Gothenburg University, Gothenburg, Sweden, [9]Sleep Disorders Centre, Department of Pulmonary Medicine, Sahlgrenska University Hospital, Gothenburg, Sweden, [10]Department of Clinical Neuroscience, Karolinska Institutet, Stockholm, Sweden, [11]Department of Neurology, Karolinska University Hospital, Stockholm, Sweden, [12]Department of Research and Development, Stockholms Sjukhem Foundation, Stockholm, Sweden, [13]Department of Oncology-Pathology, Karolinska Institutet, Stockholm, Sweden, [14]Department of Medical Sciences, Neurology, Uppsala University, Uppsala, Sweden, and [15]Department of Respiratory Medicine, Faculty of Medicine and Health, Örebro University, Örebro, Sweden

Abstract

Background: Home mechanical ventilation (HMV), including noninvasive ventilation (NIV) and invasive ventilation (IV), is increasingly used in people with amyotrophic lateral sclerosis (pwALS), but its impact on end-of-life care remains unclear. *Objective:* To examine whether ventilation pathway (NIV or IV) influences symptom prevalence and relief during the final week of life in pwALS. Secondary objectives were to assess end-of-life discussions, place and cause of death, and anticipated death. *Methods:* This national, population-based retrospective cohort study included pwALS using HMV in the Swedish National Quality Registry for Respiratory Failure (Swedevox), cross-linked with the Swedish Register of Palliative Care, which contains staff-reported end-of-life data, and other national registries. *Results:* Among 652 pwALS, 609 received NIV (mean age 66 years, 42% female) and 43 IV (mean age 60 years, 42% female). Anxiety and breathlessness were more common in the NIV group (76% vs 48% and 72% vs 31%, both $p < 0.001$), while prevalence and relief of other symptoms were similar between groups. Most patients had end-of-life discussions (NIV 87%, IV 92%). NIV patients more often died in specialized palliative wards (22% vs 7%, $p = 0.023$). Most deaths were due to ALS (96%) and were anticipated (NIV 94%, IV 88%). *Conclusions:* In pwALS with HVM, NIV was associated with higher rates of anxiety and breathlessness in the final week of life, but the ventilation pathway did not influence overall symptom relief. End-of-life communications and cause of death were similar; place of death differed by ventilation pathway; however, most deaths were anticipated.

Keywords: *amyotrophic lateral sclerosis; home mechanical ventilation; end-of-life care; invasive ventilation; non-invasive ventilation*

Correspondence: Andreas Palm, Department of Medical Sciences, Respiratory, Allergy and Sleep Research, Uppsala University, Box 256, SE-751 05, Uppsala, Sweden. E-mail: andreas.palm@medsci.uu.se
#Shared first authorship

Supplemental data for this article can be accessed online at https://doi.org/10.1080/21678421.2026.2747563.

(Received 13 July 2026; revised 25 August 2026; accepted 6 October 2026)

ISSN 2167-8421 print/ISSN 2167-9223 online © 2026 The Author(s). Published by Informa UK Limited, trading as Taylor & Francis Group.
This is an Open Access article distributed under the terms of the Creative Commons Attribution License (http://creativecommons.org/licenses/by/4.0/), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited. The terms on which this article has been published allow the posting of the Accepted Manuscript in a repository by the author(s) or with their consent.
DOI: 10.1080/21678421.2026.2747563

Introduction

Amyotrophic lateral sclerosis (ALS) is a progressive and incurable motor neuron disease characterized by degeneration of upper and lower motor neurons, leading to progressive muscle weakness, disability, and premature death (1). Following diagnosis, the approximate time of survival is two to four years (1). Early manifestations include focal muscle weakness and atrophy, eventually progressing to generalized paralysis. Respiratory muscle involvement is a central feature of disease progression; approximately 80% of people with ALS (pwALS) develop breathlessness and hypoventilation (2). Home mechanical ventilation (HMV), delivered either as noninvasive ventilation (NIV) or invasive ventilation via tracheostomy (IV), prolongs survival and at least initially improves quality of life in selected patients (3,4). Over the past three decades, the use of both NIV and IV has increased in Sweden (5). While these interventions prolong survival, they can also shape how the final phase of life unfolds, influence the dying process, the symptom burden before death, and the organization of end-of-life care.

Early integration of palliative care alongside life-prolonging interventions may facilitate symptom management, advance care planning, and support for patients and caregivers (6–8). Despite these benefits, integration of palliative care into ALS management remains variable. Although symptom control in ALS is generally considered effective, a substantial proportion of patients experience distressing symptoms such as pain, breathlessness, confusion or anxiety that are only partially relieve (9). In addition to respiratory failure, pneumonia and cardiac conditions contribute to mortality. Patients may die at home, in specialized palliative care units, or in hospital settings, sometimes following rapid clinical deterioration (9).

Despite increasing use of HMV, limited knowledge exists regarding how different ventilation pathways influence symptom burden, symptom relief, communication about end-of-life issues, and care circumstances during the final phase of life. IV is associated with substantially longer survival than NIV (10), resulting in a different disease trajectory. However, whether these differences translate into differences in symptom burden, symptom management, or end-of-life care remains unknown.

The primary aim of this study was to compare end-of-life care for Swedish pwALS treated with NIV or IV at home in the final week of life. We focused on the prevalence and relief of symptoms, including pain, anxiety, nausea, confusion, breathlessness, and terminal respiratory secretions by each treatment pathway. Secondly, we aimed to examine the presence of end-of-life (EOL) conversations, cause and place of death, and whether death was anticipated. We hypothesized that the distinct disease trajectories and models of care associated with the two ventilation pathways (NIV and IV) would result in differences in end-of-life care.

Methods

Study design and population

This retrospective cohort study was based on the national, population-based Course of DIsease in patients reported to the Swedish CPAP, Oxygen and VEntilatory RegistrY (DISCOVERY) cohort (11). We included patients with ALS (pwALS) using HMV who were reported to the National Quality Registry for Respiratory Failure (Swedevox) between January 1, 1996 and October 13, 2022. Data were cross-linked with the Swedish Register of Palliative Care (SRPC) (12), the National Cause of Death Registry (13), and the National Patient Registry (NPR) which includes physician-assigned diagnoses from public inpatient and outpatient care (14). The study was approved by the Regional Ethics Review Board at Lund University (Dnr 2018-51, and is reported in accordance with the Strengthening the Reporting of Observational Studies in Epidemiology (STROBE) guidelines (15), and the protocol is provided as Supplementary material.

Assessments

Baseline data from HMV start, demographic data (age, sex) and initial HMV pathway (NIV and IV) were retrieved from Swedevox. Transitions from noninvasive ventilation (NIV) to invasive ventilation (IV) were approximated from the reporting of procedure codes for tracheostomy in the NPR (GBA00, GBB00, GBB03, GBB96, DG020, DG024, Z430, Z930) and this group was merged with the IV-group (10).

Information on comorbid lung disease and cancer was retrieved from the NPR, defined by the International Classification of Diseases version 10 (ICD-10) codes J46-51 and C00-99, respectively.

Date and cause of death were obtained from the National Cause of Death Registry (13). Underlying cause of death was categorized as "ALS or other neurodegenerative disease" or "Other."

Palliative care data for the last week of life were entered by health care personnel on the date of death into the SRPC (12), from which we retrieved information about age at death, comorbidities, symptom prevalence and relief, EOL conversations, place of death, and whether death was expected. The following symptoms were analyzed: pain, anxiety, nausea, confusion, breathlessness, and terminal respiratory secretions (TRS). Symptom relief at some point in the final week was registered among those reporting respective symptom, and categorized as "Completely,"

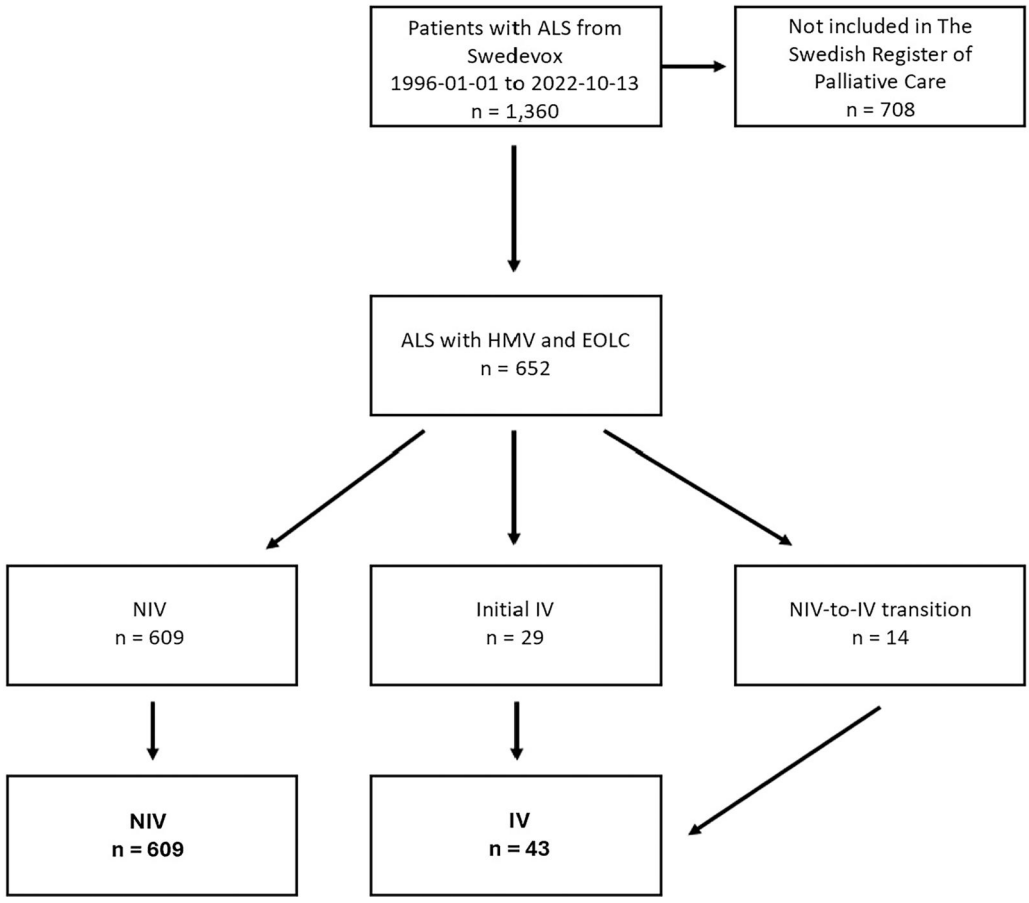

Figure 1. Flowchart of the study population. ALS: amyotrophic lateral sclerosis. HMV: home mechanical ventilation. EOLC: End-of-life care. NIV: noninvasive ventilation. IV: invasive ventilation.

"Partially," or "Not at all." EOL conversations were recorded as a binary variable (yes/no). Place of death categories were categorized as follows: home (including specialized palliative home healthcare, home healthcare, and home care), hospital (non-specialized ward/clinic/ICU), specialized palliative ward (inpatient unit specifically dedicated to specialized palliative care), and other (short-term stay, sheltered housing/care home/Residential care under the Act Concerning Support and Service for Persons with Certain Functional Impairments (LSS) (16), or other). Expected death was analyzed as a binary variable (yes/no). Approximately 56% of all annual deaths in Sweden in 2021 were included in the registry (17).

Statistical analyses

Categorical variables are presented as frequencies and percentages, and continuous variables as mean ± standard deviation (SD). Group differences between pwALS on NIV and IV regarding the assessments above were analyzed using chi-square and t-tests. When the expected cell count was less than 5, Fisher's exact test was used. A two-sided p-value < 0.05 was considered statistically significant. Statistical analyses were performed using STATA version 18.0 (StataCorp LLC, College Station, TX, USA).

Results

Study population

Of 1,360 pwALS (39% women) with HMV in Swedevox, 652 (48%) had been reported to SRPC and were included in the analyses; 609 patients with NIV and 43 with IV (Figure 1).

Compared with the NIV group, IV pwALS were approximately 6 years younger, and the duration of therapy was around 3 times longer (Table 1).

Symptoms

During the last week of life, the prevalence of anxiety and breathlessness was higher in the NIV patient group compared to the IV-group, while differences in pain, nausea, confusion and TRS were not statistically significant (Table 2).

Symptom relief

No significant differences were observed regarding symptom relief of pain, anxiety, nausea, confusion, breathlessness, or TRS between the HMV groups

Table 1. Characteristics of patients in the SRPC.

	NIV (N = 609)	IV (N = 43)
Females, (%)	253 (42)	18 (42)
Age at death (years ± SD)	67 (10)	60 (10)
Time until death since HMV start (months ± SD)	15 (21)	41 (33)
Cerebrovascular disease, (%)	89 (15)	12 (28)
Cancer, (%)	17 (3)	2 (5)

NIV: noninvasive ventilation. IV: invasive ventilation. HMV: home mechanical ventilation. EOLC: end-of-life care.

Table 2. Prevalence of six symptoms among people in the SRPC with ALS on home mechanical ventilation in their final week of life stratified by treatment group.

	NIV (N = 609)	IV (N = 43)	*p*-value	Missing data, **%**
Pain	270 (50%)	13 (36%)	0.100	12%
Anxiety	402 (76%)	16 (49%)	**<0.001**	14%
Nausea	42 (8%)	4 (12%)	0.41	15%
Confusion	44 (8%)	2 (7%)	0.71	14%
Breathlessness	393 (72%)	11 (31%)	**<0.001**	11%
Terminal Respiratory Secretions	302 (54%)	15 (41%)	0.10	9%

NIV: noninvasive ventilation. IV: invasive ventilation.

(Figure 2). Most patients experienced improvement in symptoms.

End-of-life conversation

The frequency of documented end-of-life conversations was similar between groups: 87% ($n = 458$) in the NIV group compared to 92% ($n = 33$) in the IV group ($p = 0.452$).

Place and cause of death

Fewer in the IV-group died in specialized palliative wards, while a similar proportion died at home compared with the NIV group (Table 3). The main underlying cause of death for the vast majority of pwALS was neurological disease (ALS), accounting for 96% of cases in both groups ($p = 0.905$). There was one recorded suicide in the NIV group; no further information on the circumstances was available. Most deaths were classified as expected in both groups, 94% in the NIV group and 88% in the IV group ($p = 0.181$).

Discussion

Main findings

This large national, population-based study of Swedish people with ALS treated with HMV identified three important findings in end-of-life care. First, pwALS treated with IV reported less breathlessness and anxiety during the last week of life compared to patients with NIV. Second, ventilation pathway was associated with differences in place of death, with fewer IV-treated patients dying in specialized palliative wards. Third, overall symptom relief, end-of-life conversations, cause of death, and expected death were similar between the IV and NIV groups.

Interpretation and relevance

The lower prevalence of breathlessness and anxiety in the IV group, despite comparable overall symptom relief between groups, is clinically plausible. Although NIV alleviates hypoventilation and prolongs survival in ALS (3), treatment success depends on mask tolerance, secretion management, prevention of air leaks and patient–ventilator synchrony. Ineffective NIV may contribute to persistent air hunger and respiratory discomfort (18). In contrast, IV via tracheostomy bypasses the upper airway and may provide more stable ventilatory support and facilitate secretion management, which could partly explain the lower prevalence of dyspnea observed in the IV group. The lower prevalence of anxiety in the IV group should be interpreted cautiously, as symptoms were staff-reported and communication impairment in advanced ALS may lead to under-recognition of subjective distress. However, long-term IV in the Swedish home-care setting does not generally involve sedation or reduced consciousness. The substantially longer duration of ventilatory treatment in the IV group also indicates different disease trajectories between the groups. As information on functional and cognitive status was unavailable, we could not determine whether

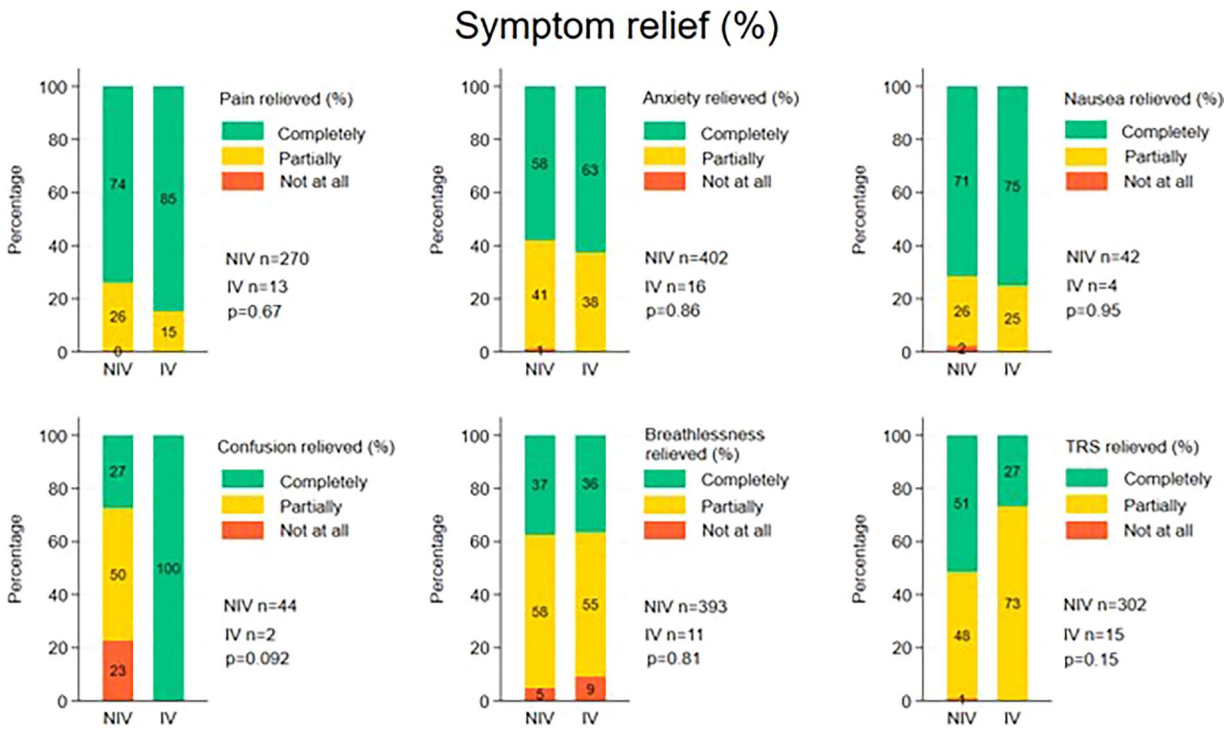

Figure 2. Symptom relief in the last week of life in pwALS in the SRPC on NIV or IV. NIV: noninvasive ventilation, IV: invasive ventilation, TRS: terminal respiratory secretions

Table 3. Place of death of patients in the SRPC presented per treatment group.

	NIV (N = 609)	IV (N = 43)	*p*-value
Home	238 (39%)	20 (47%)	0.703
Hospital	137 (22%)	10 (23%)	0.525
Specialized palliative ward	132 (22%)	3 (7%)	**0.023**
Other location	102 (17%)	10 (23%)	–

Analyses of group differences made between each place of death compared with other location.
NIV: noninvasive ventilation, IV: invasive ventilation.

differences in disease severity or communication ability contributed to differences in symptom detection or management. Nevertheless, the higher prevalence of anxiety in the NIV group may partly reflect the higher prevalence of dyspnea, given the close relationship between breathlessness and anxiety (19). The bidirectional relationship between dyspnea and anxiety may therefore contribute to greater symptom burden among NIV-treated pwALS.

Importantly, these differences in symptom prevalence did not result in differences in documented symptom relief between ventilation pathways. Most patients experienced partial or complete relief, suggesting that symptom management in the final week of life was generally effective once symptoms were recognized. This is consistent with previous SRPC-based data showing that most individual end-of-life symptoms in ALS were either absent or completely relieved, although approximately half of patients experienced at least one incompletely relieved symptom (9). The present study extends these findings by distinguishing between symptom prevalence and symptom relief according to ventilation pathway. It also aligns with broader reviews of end-of-life care in ALS emphasizing the effectiveness of multidisciplinary palliative approaches in alleviating distress and supporting quality of life alongside life-prolonging treatments (20).

End-of-life conversations were documented for approximately 90% of patients in both groups, indicating that end-of-life issues were commonly addressed, consistent with recommendations for early and repeated advance care planning in ALS (9,21,22). However, the registry does not provide information on the content, quality, timing, or emotional support associated with these conversations. Notably, most deaths were classified as expected, indicating that the terminal phase was often recognized in pwALS reported to SRPC in time to allow planning and symptom management. Taken together, these findings support combining life-prolonging respiratory support with timely palliative care throughout the disease course (20).

Overall, approximately 40% of patients died at home and 20% in hospital, proportions that are the reverse of those reported in neighboring Norway despite broadly similar healthcare systems (23). This may reflect differences in organization of community-based ALS and palliative care services, availability of specialized home care, and local traditions regarding end-of-life management. Place

of death differed significantly between groups, with IV-treated patients less likely to die in specialized palliative wards and more likely to die at home. This difference may partly reflect access to specialized inpatient palliative care, as caring for patients receiving IV requires appropriate expertise and resources for tracheostomy ventilation. Patients treated with IV often have established, resource-intensive home-care arrangements, which may facilitate anticipatory planning and enable death in the home setting. Transfer to inpatient palliative units may also be logistically more complex for these patients. Overall, ventilation pathways may shape the organization and location of end-of-life care more than the adequacy of symptom control, emphasizing the importance of aligning care planning with patient preferences and clinical trajectories.

Neurological disease was the primary recorded cause of death in most patients in both groups, which is consistent with the low comorbidity burden observed in this cohort. Patients with ALS commonly die from respiratory complications that arise as a direct consequence of the underlying neurodegenerative disease, and our findings are in line with previous studies (9,24).

Strengths and limitations

Strengths of this study include the nationwide population-based design, linkage of validated Swedish registries, and focus on clinically relevant end-of-life outcomes (12,25). Several limitations should be acknowledged. The retrospective observational design precludes causal inference, and residual confounding is likely, as patients selected for IV were younger and may have differed in phenotype, social support, or disease trajectory. The relatively small IV cohort limits statistical precision. Furthermore, symptoms were staff-reported rather than patient-reported, and subjective symptoms such as anxiety may therefore have been under-recognized, particularly in patients with impaired communication. Similarly, data on end-of-life conversations captured their occurrence but not their content, quality, timing, or associated emotional support. Detailed measures of disease severity, functional status, cognitive impairment, and communication ability were unavailable, limiting our ability to assess how differences in disease trajectory may have influenced symptom detection and management. Some variables also had missing data. In addition, only approximately half of patients with ALS receiving HMV were represented in the SRPC, which may have introduced selection bias. However, few differences were seen between the non-SRPC and SRPC patients (Supplemental Table 1), suggesting that our results are generalizable to the broader population of pwALS receiving HMV. Nonetheless, these findings provide novel real-world evidence that ventilation pathway may shape symptom experience and care setting at the end of life in ALS.

Clinical implications

These findings have several clinical implications for ALS care. Patients treated with NIV may require particularly close monitoring for persistent breathlessness and anxiety near the end of life, including assessment of mask tolerance, air leaks, secretion burden, and patient-ventilator synchrony. Unresolved symptoms should prompt optimization of ventilatory support alongside timely palliative interventions, including pharmacological and non-pharmacological measures for dyspnea and distress. The high proportion of anticipated deaths and end-of-life discussions supports early, repeated advance care planning. The high proportion of home deaths, especially among IV-treated patients, highlights the need for coordinated community care, caregiver support, and access to specialist palliative services. Respiratory support and palliative care should be integrated throughout the disease course. Patients receiving IV may benefit from more experienced home-care support, reflecting the specialized training required to manage the treatment. Strengthening symptom-management competence in home care for patients using NIV could potentially reduce end-of-life distress and support care at home.

Conclusion

In this national population-based study of pwALS receiving HMV, NIV was associated with more frequently reported breathlessness and anxiety in the final week of life, while reported symptom relief and end-of-life communication were similar between NIV and IV groups. Most deaths were anticipated, although place of death differed by ventilation pathway. These findings suggest that ventilation pathway may influence symptom experience, and care setting, at the end-of-life. NIV should not be assumed to fully relieve respiratory distress near end of life – persistent breathlessness and anxiety require active reassessment, optimization of ventilatory support, and timely palliative management.

Acknowledgements

We sincerely thank the patients for sharing their experiences during the final week of life. Their contribution was invaluable to this manuscript.

Author contributions

CRediT: **Mikaela Dalkvist**: Formal analysis, Visualization, Writing – original draft, Writing –

review & editing; **Karin Ersson**: Writing – original draft, Writing – review & editing; **Stefan Bergström**: Writing – review & editing; **Magnus Ekström**: Writing – review & editing; **Össur Ingi Emilsson**: Writing – review & editing; **Ludger Grote**: Writing – review & editing; **Caroline Ingre**: Writing – review & editing; **Ulf Kläppe**: Writing – review & editing; **Staffan Lundström**: Writing – review & editing; **Ingela Nygren**: Writing – review & editing; **Josefin Sundh**: Writing – review & editing; **Andreas Palm**: Conceptualization, Formal analysis, Funding acquisition, Methodology, Resources, Supervision, Writing – review & editing.

Declaration of interest

The authors declare no conflict of interest related to this work. Outside the topic of the current study, AP has received personal fees for lectures and educational activities from ResMed, Breas, and Chiesi.

Funding

AP was supported by the Swedish Heart and Lung Foundation (Hjärt-Lungfonden) (grants 20230392 and 20251024), the Swedish ALS Foundation (Insamlingsstiftelsen ALS-fonden) (grant 2024.1), Björklund's foundation (Björklunds fond) (grant SLS-1016320), ALF (the agreement concerning the research and education of doctors) (ALF-979044) and the Swedish Respiratory Society. ME was supported by an unrestricted grant from the Swedish Research Council (Vetenskapsrådet, Dnr: 2019–02081).

Guarantor statement

AP take responsibility for the content of the manuscript, including the data and analysis.

Use of artificial intelligence

Generative artificial intelligence (ChatGPT, GPT-5.5 Thinking, OpenAI) was used solely to support language editing and improve the clarity and readability of the manuscript. All analyses, interpretation of findings, and scientific conclusions were conducted by the authors, who take full responsibility for the content of the manuscript.

ORCID

Karin Ersson http://orcid.org/0000-0003-3185-3316
Magnus Ekström http://orcid.org/0000-0002-7227-5113
Ludger Grote http://orcid.org/0000-0002-7405-1682
Caroline Ingre http://orcid.org/0000-0001-5327-7204
Ulf Kläppe http://orcid.org/0000-0001-5832-038X
Staffan Lundström http://orcid.org/0000-0002-5699-7757
Ingela Nygren http://orcid.org/0000-0002-0091-924X
Josefin Sundh http://orcid.org/0000-0003-1926-8464
Andreas Palm http://orcid.org/0000-0002-0590-0417

Data availability statement

The data that support the findings of this study were obtained from the Swedish National Quality Registry for Respiratory Failure (Swedevox), the Swedish Register of Palliative Care (SRPC), the National Patient Register, and the Cause of Death Register. The data are not publicly available due to Swedish data protection legislation but may be accessed by qualified researchers following approval from the Swedish Ethical Review Authority and the respective registry holders.

References

1. Brown RH, Al-Chalabi A. Amyotrophic lateral sclerosis. N Engl J Med. 2017;377:1602.
2. Vender RL, Mauger D, Walsh S, Alam S, Simmons Z. Respiratory systems abnormalities and clinical milestones for patients with amyotrophic lateral sclerosis with emphasis upon survival. Amyotroph Lateral Scler. 2007;8:36–41.
3. Bourke SC, Bullock RE, Williams TL, Shaw PJ, Gibson GJ. Noninvasive ventilation in ALS: indications and effect on quality of life. Neurology. 2003;61:171–7.
4. Hayashi N, Atsuta N, Yokoi D, Nakamura R, Nakatochi M, Katsuno M, et al. Prognosis of amyotrophic lateral sclerosis patients undergoing tracheostomy invasive ventilation therapy in Japan. J Neurol Neurosurg Psychiatry. 2020;91:285–90.
5. Palm A, Grote L, Einarsson J, Hansson D, Ljunggren M, Sundh J, et al. Evolution of home mechanical ventilation in Sweden over 27 years: changing trends in incidence, prevalence, and patient characteristics - The population-based the course of Disease in Patients Reported to the Swedish CPAP Oxygen and Ventilator Registry (DISCOVERY) Study. CHEST Pulm. 2024;2:100108.
6. Fahrner-Scott K, Zapata C, O'Riordan DL, Cohen E, Rosow L, Pantilat SZ, et al. Embedded palliative care for amyotrophic lateral sclerosis: a pilot program and lessons learned. Neurol Clin Pract. 2022;12:68–75.
7. Zwicker J, Smith IC, Rice J, Murphy R, Breiner A, McNeely S, et al. Palliative care at any stage of amyotrophic lateral sclerosis: a prospective feasibility study. Front Med (Lausanne). 2023;volume10:1204816. 2023.
8. ALS Kunskapsstöd 2026 [3 July 2026]. Available from: https://vardpersonal.1177.se/Stockholm/kunskapsstod/vardforlopp/als/.

9. Sennfält S, Kläppe U, Thams S, Samuelsson K, Press R, Fang F, et al. Dying from ALS in Sweden: clinical status, setting, and symptoms. Amyotroph Lateral Scler Frontotemporal Degener. 2023;24:237–45.
10. Palm A, Ekström M, Einarsson J, Ersson K, Ingre C, Kläppe U, et al. Home mechanical ventilation for amyotrophic lateral sclerosis—a national study of survival and treatment trends across 27 years. Ann Am Thorac Soc. 2026;23:aaoag219.
11. Palm A, Ågren K, Grote L, Ljunggren M, Midgren B, Sundh J, et al. Course of DISease In patients reported to the Swedish CPAP Oxygen and VEntilator RegistrY (DISCOVERY) with population-based controls. BMJ Open. 2020;10:e040396.
12. Martinsson L, Heedman PA, Lundström S, Axelsson B. Improved data validity in the Swedish Register of Palliative Care. PLoS One. 2017;12:e0186804.
13. Brooke HL, Talbäck M, Hörnblad J, Johansson LA, Ludvigsson JF, Druid H, et al. The Swedish cause of death register. Eur J Epidemiol. 2017;32:765–73.
14. Everhov ÅH, Frisell T, Osooli M, Brooke HL, Carlsen HK, Modig K, et al. Diagnostic accuracy in the Swedish national patient register: a review including diagnoses in the outpatient register. Eur J Epidemiol. 2025;40:359–69.
15. von Elm E, Altman DG, Egger M, Pocock SJ, Gøtzsche PC, Vandenbroucke JP, STROBE Initiative. The Strengthening the Reporting of Observational Studies in Epidemiology (STROBE) statement: guidelines for reporting observational studies. Lancet. 2007;370:1453–7.
16. Socialstyrelsen. National Register of Municipal Support and Service for Persons with Certain Functional Impairments: Socialstyrelsen; 2023 [cited 2026 April 27]. Available from: https://www.socialstyrelsen.se/en/statistics-and-data/registers/national-register-of-municipal-support-and-service-for-persons-with-certain-functional-impairments/.
17. Årsrapport Pallitiva registret 2021 [Available from: www.palliativregistret.se.
18. Sancho J, Ferrer S, Signes-Costa J. Noninvasive ventilation effectiveness in amyotrophic lateral sclerosis. J Clin Med. 2025;14:8609.
19. Sher Y, Desai N, Sole J, D'Souza MP. Dyspnea and dyspnea-associated anxiety in the ICU Patient population: a narrative review for CL psychiatrists. J Acad Consult Liaison Psychiatry. 2024;65:54–65.
20. Connolly S, Galvin M, Hardiman O. End-of-life management in patients with amyotrophic lateral sclerosis. Lancet Neurol. 2015;14:435–42.
21. Khan A, Frazer-Green L, Amin R, Wolfe L, Faulkner G, Casey K, et al. Respiratory management of patients with neuromuscular weakness: An American College of Chest Physicians Clinical Practice Guideline and Expert Panel Report. Chest. 2023;164:394–413.
22. Seeber AA, Pols AJ, Hijdra A, Grupstra HF, Willems DL, de Visser M. Advance care planning in progressive neurological diseases: lessons from ALS. BMC Palliat Care. 2019;18:50.
23. Johansen H, Nakken O, Holmøy T, Fredheim OMS. Disease trajectories and end of life care in a Norwegian ALS cohort. BMC Neurol. 2026;26:155.
24. Larson TC, Goutman SA, Davis B, Bove FJ, Thakur N, Mehta P. Causes of death among US decedents with ALS: An eye toward delaying mortality. Ann Clin Transl Neurol. 2023;10:757–64.
25. Ekström M, Albrecht D, Andersson S, Grote L, Kärrsten Rundström B, Palm A, et al. Validation of the Swedevox registry of continuous positive airway pressure, long-term mechanical ventilator and long-term oxygen therapy. ERJ Open Res. 2021;7:00340–2020.