Tidsskriftet Tidsskrift for Den norske legeforening

Ventilation of patients with amyotrophic lateral sclerosis

PERSPECTIVES

OLE-BJØRN TYSNES

E-mail: obty@haukeland.no
Ole-Bjørn Tysnes, specialist in neurology, senior consultant at the Department of Neurology, Haukeland University Hospital and adjunct professor at the Department of Clinical Medicine, University of Bergen.
The author has completed the ICMJE form and declares no conflicts of interest.

TRYGVE HOLMØY

Trygve Holmøy, specialist in neurology, senior consultant and head of section for neuroimmunological diseases at the Department of Neurology, Akershus University Hospital, and adjunct professor at the Institute of Clinical Medicine, University of Oslo.
The author has completed the ICMJE form and declares no conflicts of interest.

SOLFRID INDREKVAM

Solfrid Indrekvam, specialist in pulmonary diseases and senior consultant at the Thoracic Department, Haukeland University Hospital, and head of the Norwegian Register for Long-term Mechanical Ventilation.
The author has completed the ICMJE form and declares no conflicts of interest.

OVE FONDENÆS

Ove Fondenæs, specialist in pulmonary diseases and senior consultant at the Thoracic Department, Haukeland University Hospital.
The author has completed the ICMJE form and declares no conflicts of interest.

Just under 10 % of patients with amyotrophic lateral sclerosis receive life-prolonging ventilation via a tracheostomy. There is no international consensus on the planning of life-prolonging invasive ventilation for patients with this disease.

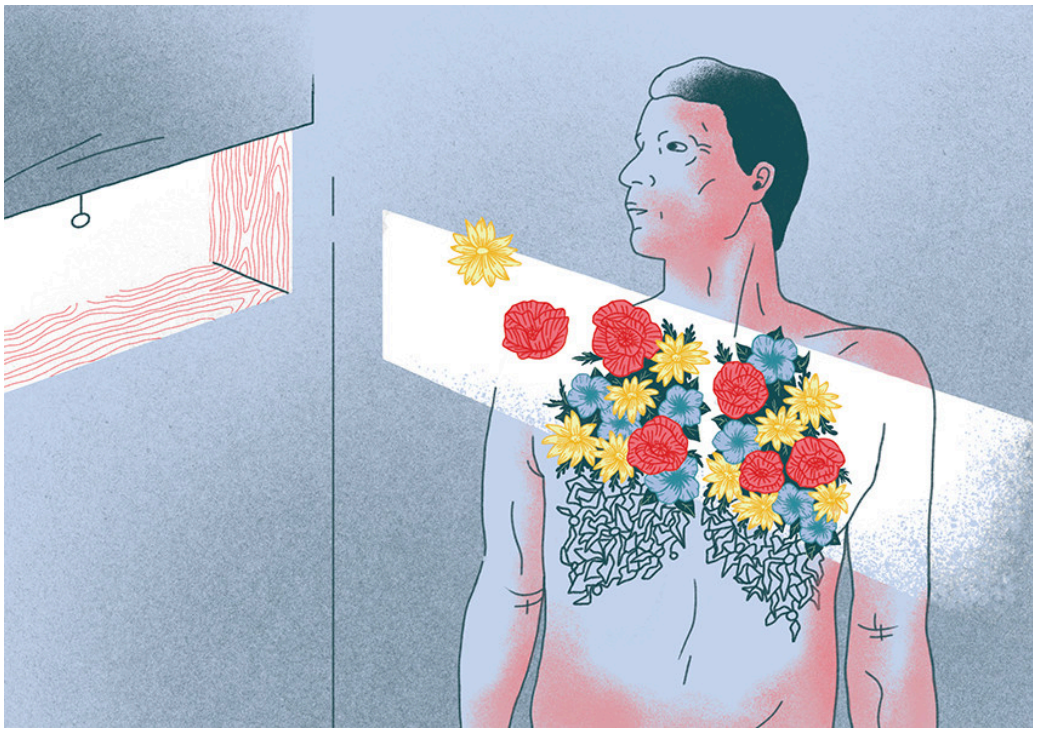

Illustration: Frida Strømme

The annual incidence rate of amyotrophic lateral sclerosis (ALS) in Norway is approximately 3 per 100 000, and the incidence is slightly higher for men than women (1, 2). About 400 people are living with the disease. The average life expectancy following diagnosis is two years (3), and approximately 10 % of patients live more than ten years. By far the most common cause of death is respiratory failure. Although ALS is considered to be a motor disease, it is well known that around half of patients develop a frontotemporal mild cognitive impairment, and up to 25 % develop dementia. This typically manifests itself in the patient's reduced awareness of the disease or a language disorder (4).

ALS patients should be treated as part of a close collaboration between neurologists and pulmonologists. Current practice is that patients who develop incipient respiratory impairment will be offered non-invasive ventilation (a home ventilator with a mask). Symptoms of hypoventilation may include headaches and poorer general health or daytime fatigue. Non-invasive ventilation improves these symptoms and has also been shown to prolong life (5, 6), but technical issues can arise for patients with bulbar symptoms such as weakness of the tongue and dysphagia. Ventilation via a tracheostomy may be suitable for these patients and in patients who specifically want life-prolonging treatment. However, this method presents an ethical dilemma for the patient, their family and healthcare personnel (7).

What does the law say?

The Patients' Rights Act aims to ensure equality of access and quality in primary care and specialist health services. In 2012, the *National Guide for Long-Term Mechanical Ventilation* was published in Norway (8). The guide stipulates that the specialist health service is responsible for deciding whether mechanical ventilation should be initiated in individual patients, but that the patient and their family must have the opportunity to contribute to the decision-making.

The patient cannot therefore *demand* life-prolonging ventilation via a tracheostomy. The final decision must be made by the consultant, usually after assessment and discussion in the interdisciplinary ALS team Box 1). However, the patient and their family often exert considerable pressure on the doctor. There are several examples of patients pressuring doctors into changing decisions on life-prolonging treatment ((9). There is no established legal practice.

Box 1 Before providing life-prolonging long-term mechanical ventilation for ALS, the authors recommend that the following are considered/discussed:

The patient is considered to be cognitively intact by experienced clinicians.

The patient is able to use appropriate communication aids.

Non-invasive ventilation (BiPAP/home ventilator with a mask) does not meet the patient's ventilation needs.

Both the patient and their family have been informed about the expected progression of the disease and the challenges of ventilation via a tracheostomy.

The patients must be consistent in their wish for treatment. Information about the treatment must be given several times.

The patient must be informed about alternatives for palliative care.

The patient's motivation for choosing a tracheostomy must be clarified.

Consideration must have been given to whether resources can be established that meet the patient's care needs following invasive ventilation.

The criteria and conditions for terminating ventilator support must be discussed with the patient and their family.

How typical is this intervention?

The use of ventilation via a tracheostomy in ALS patients varies considerably, and the intervention is seldom used in England. It is most common in Japan, where up to 45 % of patients receive life-prolonging treatment. In Europe and

the United States, use varies from 5 % to 10 % (6, 10).

In the period 2002–07, 6.7 % of men and 3.8 % of women with ALS in Norway were given ventilation via tracheostomy (11). Data from the Norwegian Register for Long-term Mechanical Ventilation (LTMV register) show that in 2017, 113 patients received active treatment in the form of long-term mechanical ventilation, 32 (28 %) underwent a tracheostomy and 81 (72 %) were given non-invasive respiratory support. In the period 2015–20, long-term mechanical ventilation was initiated for 256 ALS patients in Norway (12).

Ethical considerations

Both non-invasive and invasive ventilation are initiated with the aim of improving the patient's quality of life, and in some cases of prolonging their life. Fatigue, headaches and a sensation of choking on obstructions in the airways are symptoms that reduce the quality of life in the advanced stages of ALS. Ventilation via a tracheostomy will enable patients to live for many years. A Danish study showed that tracheostomy patients received ventilation for 2.8–4.7 years (13). Prolonged survival of 2–4 years using invasive ventilation has also been demonstrated (14, 15). A recent Japanese study showed an average prolonged life of 6.7 years (16).

> ***«It is difficult for patients to assess quality of life when the alternative is death»***

Multiple studies show that ALS patients maintain a good quality of life with this type of advanced treatment, and that patients often rate their own quality of life higher than the doctors (15, 16, 17). Psychological, social and existential factors have a greater impact on self-reported quality of life in ALS patients than physical function level (18). In practice, however, it is often very difficult to assess the quality of life in patients showing signs of locked-in syndrome, i.e. conscious but unable to speak or move. This is particularly the case if cognition and language function are also affected. It is difficult for patients to assess quality of life when the alternative is death (7). Patients may feel guilt towards family members for the situation they are all in when a patient is receiving ventilation via a tracheostomy.

One of the challenges when ALS patients are receiving ventilation via a tracheostomy is that a decision often has to be made about whether to withdraw ventilation (19). In our view, patients must be informed of this before the intervention is offered. Patients will have the right to discontinue the ventilation whenever they wish, but this can of course be a very difficult decision. A significant challenge for patients and therapists is the patient's reduced ability to communicate. Prior to initiating the intervention, an agreement will normally be made with the patient to switch off the ventilator when it is no longer possible for the therapist and patient to communicate. However, agreements of this nature are not legally binding for the patient, who will subsequently have the opportunity to retract it. However, the agreement

will help to ensure that the patient is informed and has reflected on these matters, and will serve as a record of the patient's wishes when ventilation was initiated. In the final phase, eye communication will be difficult. The therapist may be afraid of not fully understanding what the patient means. This can lead to over-treatment.

> ***«Negative consequences for the family must be considered when deciding whether to provide ventilation»***

An important principle in medical ethics is that no harm comes to the patient. In ALS, however, there is also the potential for harming the family. Ventilation via a tracheostomy affects not only the patient but also the family and healthcare personnel. Cultural differences may dictate whether a family is able to accept invasive ventilation (20). Negative consequences for the family must be considered when deciding whether to provide ventilation. We believe that this must be discussed with the patient prior to initiating ventilation via a tracheostomy. There is very little research on the strain experienced by patients' families, but it has been shown that they report a reduced quality of life to a greater extent than the patients (21) and that they are faced with a major burden (20, 22). Spouses can feel trapped, and divorce or separation can be regarded as ruthless. Patients' families therefore have no choice but to stick with the patient. The time perspective is uncertain, and this has a considerable effect on their quality of life. In a US study, more than half of the family members were depressed when the decision was made to provide ventilation, but the number who had symptoms of depression fell during the intervention period (23). The patient cannot be solely responsible for this type of decision. Discussion between the doctor, patient and family is essential for shedding light on how ventilation via a tracheostomy will affect the patient and their family.

Another important principle in all medical interventions is respect for the patient's autonomy.

ALS patients may experience impaired cognitive function, ranging from a subtle language disorder to impaired executive functions (24). We believe that consideration should always be given to whether patients understand what the decisions on future interventions may mean for the family and other carers. This is especially important for patients with mild frontal impairment without clinical frontotemporal dementia. These patients will lack understanding of how the disease and treatment may impact on their spouse and children. A decision must be made on whether the patient has sufficient awareness to understand the consequences of decisions. If the cognitive impairment affects language function, it will be very difficult to communicate with patients receiving invasive ventilation.

> ***«'Choosing to die' is a hard choice for patients, but so is 'choosing to live'»***

In our experience, many patients 'avoid the decision' on life-prolonging ventilation. Talking about it is stressful for them, and they are very aware that it is also extremely difficult for their family. 'Choosing to die' is a hard choice for patients, but so is 'choosing to live'. The patient's wish to avoid this discussion must be respected, even if it is difficult for therapists of patients in an advanced stage that no decision is made on life-prolonging treatment. In our opinion, ALS patients who do not want to talk about life-prolonging treatment are not suitable for ventilation via a tracheostomy.

Conclusion

Amyotrophic lateral sclerosis is one of the few diseases in adults where it is possible to prolong the patient's life after the disease has reached the final stage where it is no longer possible to survive without advanced medical interventions. Ventilation via a tracheostomy is performed in approximately 6 % of ALS patients in Norway. The decision is made by a doctor in consultation with the patient, the patient's family and other medical personnel. It is our recommendation that patients and their families are thoroughly briefed on the situation. The patient must be suitable for ventilation in terms of their wishes and cognitive function, the family and primary health service must be able to provide the necessary care and a plan must be in place for when ventilation should be withdrawn.

LITERATURE

1. Nakken O, Lindstrøm JC, Tysnes OB et al. Assessing amyotrophic lateral sclerosis prevalence in Norway from 2009 to 2015 from compulsory nationwide health registers. Amyotroph Lateral Scler Frontotemporal Degener 2018; 19: 303–10. [PubMed][CrossRef]

2. Nakken O, Lindstrøm JC, Tysnes OB et al. Mortality trends of amyotrophic lateral sclerosis in Norway 1951-2014: an age-period-cohort study. J Neurol 2016; 263: 2378–85. [PubMed][CrossRef]

3. Rooney J, Byrne S, Heverin M et al. A multidisciplinary clinic approach improves survival in ALS: a comparative study of ALS in Ireland and Northern Ireland. J Neurol Neurosurg Psychiatry 2015; 86: 496–501. [PubMed][CrossRef]

4. Woolley SC, Strong MJ. Frontotemporal dysfunction and dementia in amyotrophic lateral sclerosis. Neurol Clin 2015; 33: 787–805. [PubMed] [CrossRef]

5. Radunovic A, Annane D, Rafiq MK et al. Mechanical ventilation for amyotrophic lateral sclerosis/motor neuron disease. Cochrane Database Syst Rev 2017; 10: CD004427. [PubMed][CrossRef]

6. Spittel S, Maier A, Kettemann D et al. Non-invasive and tracheostomy invasive ventilation in ALS: utilisation and survival rates in a cohort study over 12 years in Germany. Eur J Neurol 2020; 28: 1160–71. [PubMed] [CrossRef]

7. Magelssen M, Holmøy T, Horn MA et al. Ethical challenges in tracheostomy-assisted ventilation in amyotrophic lateral sclerosis. J Neurol 2018; 265: 2730–6. [PubMed][CrossRef]

8. Nasjonal veileder for langtids mekanisk ventillasjon. Kapittel 9.4.1. Oslo: Helsedirektoratet, 2012. https://www.helsedirektoratet.no/veiledere/langtids-mekanisk-ventilasjon-ltmv/Nasjonal-veileder-for-langtids-mekanisk-ventilasjon-ltmv.pdf/_/attachment/inline/c0c5f13b-58b9-4cef-8a2c-4e6afdf26e99:4e60148bd277c5f95bf3b244eaf1bbc764b0d6fb/Nasjonal-veileder-for-langtids-mekanisk-ventilasjon-ltmv.pdf Accessed 15.3.2021.

9. Bakke P, Barth-Heyerdahl L. Pål får pustehjelpen Freddy nektes. TV2 5.3.2013. https://www.tv2.no/a/13641565/ Accessed 15.3.2021.

10. Weber C, Fijalkowska B, Ciecwierska K et al. Existential decision-making in a fatal progressive disease: how much do legal and medical frameworks matter? BMC Palliat Care 2017; 16: 80. [PubMed][CrossRef]

11. Tollefsen E, Midgren B, Bakke P et al. Amyotrophic lateral sclerosis: gender differences in the use of mechanical ventilation. Eur J Neurol 2010; 17: 1352–7. [PubMed][CrossRef]

12. LTMV rapport. Årsrapport for 2019 med plan for forbedringstiltak. Bergen: Nasjonalt register for langtids mekanisk ventilasjon, 2020. https://www.kvalitetsregistre.no/sites/default/files/17_arsrapport_2019_ltmv.pdf Accessed 15.3.2021.

13. Dreyer P, Lorenzen CK, Schou L et al. Survival in ALS with home mechanical ventilation non-invasively and invasively: a 15-year cohort study in west Denmark. Amyotroph Lateral Scler Frontotemporal Degener 2014; 15: 62–7. [PubMed][CrossRef]

14. Spataro R, Bono V, Marchese S et al. Tracheostomy mechanical ventilation in patients with amyotrophic lateral sclerosis: clinical features and survival analysis. J Neurol Sci 2012; 323: 66–70. [PubMed][CrossRef]

15. Sanjuán-López P, Valiño-López P, Ricoy-Gabaldón J et al. Amyotrophic lateral sclerosis: impact of pulmonary follow-up and mechanical ventilation on survival. A study of 114 cases. Arch Bronconeumol 2014; 50: 509–13. [PubMed][CrossRef]

16. Hayashi N, Atsuta N, Yokoi D et al. Prognosis of amyotrophic lateral sclerosis patients undergoing tracheostomy invasive ventilation therapy in Japan. J Neurol Neurosurg Psychiatry 2020; 91: 285–90. [PubMed] [CrossRef]

17. Vianello A, Arcaro G, Palmieri A et al. Survival and quality of life after tracheostomy for acute respiratory failure in patients with amyotrophic lateral sclerosis. J Crit Care 2011; 26: 329.e7–14. [PubMed][CrossRef]

18. Chiò A, Gauthier A, Montuschi A et al. A cross sectional study on determinants of quality of life in ALS. J Neurol Neurosurg Psychiatry 2004; 75: 1597–601. [PubMed][CrossRef]

19. Holmøy T, Aarrestad S, Engstrand P et al. Avslutning av respiratorbehandling ved amyotrofisk lateral sklerose. Tidsskr Nor Legeforen 2009; 129: 628–31. [PubMed][CrossRef]

20. Kaub-Wittemer D, Steinbüchel N, Wasner M et al. Quality of life and psychosocial issues in ventilated patients with amyotrophic lateral sclerosis and their caregivers. J Pain Symptom Manage 2003; 26: 890–6. [PubMed] [CrossRef]

21. Christodoulou G, Goetz R, Ogino M et al. Opinions of Japanese and American ALS caregivers regarding tracheostomy with invasive ventilation (TIV). Amyotroph Lateral Scler Frontotemporal Degener 2015; 17: 47–54. [PubMed][CrossRef]

22. Lerum SV, Solbrække KN, Frich JC. Family caregivers' accounts of caring for a family member with motor neurone disease in Norway: a qualitative study. BMC Palliat Care 2016; 15: 22. [PubMed][CrossRef]

23. Rabkin JG, Albert SM, Rowland LP et al. How common is depression among ALS caregivers? A longitudinal study. Amyotroph Lateral Scler 2009; 10: 448–55. [PubMed][CrossRef]

24. Woolley SC, Rush BK. Considerations for clinical neuropsychological evaluation in amyotrophic lateral sclerosis. Arch Clin Neuropsychol 2017; 32: 906–16. [PubMed][CrossRef]

Publisert: 14 May 2021. Tidsskr Nor Legeforen. DOI: 10.4045/tidsskr.20.1030
Received 16.12.2020, first revision submitted 19.2.2021, accepted 15.3.2021.
Copyright: © Tidsskriftet 2026 Downloaded from tidsskriftet.no 4 June 2026.