

Is tracheostomy still an option in amyotrophic lateral sclerosis?

Reflections of a multidisciplinary workgroup

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Summary

QUESTION UNDER STUDY: Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disease with a poor prognosis. Survival and quality of life of ALS patients have improved through the implementation of multidisciplinary approaches, the use of percutaneous gastrostomy and of noninvasive (NIV) or invasive ventilation. The question of whether or not to propose invasive ventilation (by tracheostomy: TPPV) to ALS patients remains a matter of debate.

METHODS: The study reviews the medical literature, the practice in three Swiss and two large French ALS expert centres and reports the results of a workgroup on invasive ventilation in ALS.

RESULTS: Improved management of secretions and use of different interfaces allows NIV to be used 24-hours-a-day for prolonged periods, thus avoiding TPPV in many cases. TPPV is frequently initiated in emergency situations with lack of prior informed consent. TPPV appears associated with a lesser quality of life and a higher risk of institution-

alisation than NIV. The high burden placed on caregivers who manage ALS patients is a major problem with a clear impact on their quality of life.

CONCLUSIONS: Current practice in Switzerland and France tends to discourage the use of TPPV in ALS. Fear of a “locked-in syndrome”, the high burden placed on caregivers, and unmasking cognitive disorders occurring in the evolution of ALS are some of the caveats when considering TPPV. Most decisions about TPPV are taken in emergency situations in the absence of advance directives. One exception is that of young motivated patients with predominantly bulbar disease who “fail” NIV.

Key words: amyotrophic lateral sclerosis; motor neurone disease; quality of life; noninvasive ventilation; tracheostomy; invasive ventilation

Introduction

Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease characterised by loss of motor neurones in the spinal cord, brainstem, and motor cortex leading to progressive loss of motor functions, causing tetraplegia, dysphagia and dysphonia, as well as respiratory failure [1]. Annual incidence is approximately 1.5–2.5/100,000, with a prevalence of 6–8/100,000. Except for a minority of long-term survivors [2, 3], mean survival from the time of appearance of the first symptoms is 2 to 4 years. Respiratory failure is the main cause of mortality in untreated subjects.

Recent studies have shown that a multidisciplinary approach [4–7] to patients with ALS is associated with decreased hospital admissions and length of hospital stay, and increased use of ventilator support, percutaneous endoscopic gastrostomy (PEG) and palliative referral [8], when compared with usual care. One Irish study documented

Abbreviations:

AD	advance directive
ALS	amyotrophic lateral sclerosis
ARF	acute respiratory failure
HRQL	health-related quality of life
ICU	intensive care unit
IMV	invasive mechanical ventilation (= TPPV)
LTMV	long-term mechanical ventilation
MV	mechanical ventilation
NIV	noninvasive ventilation (refers essentially to nasal or facial positive pressure ventilation)
PEG	percutaneous endoscopic gastrostomy
RCU	respiratory care unit
RF	respiratory failure
TPPV	positive pressure ventilation via a tracheostomy

prolonged survival (7.5 months) [7]. Patients who benefit from a multidisciplinary approach also appear to receive more aids and appliances and have a higher quality of life [9, 10].

Among aids and appliances, appropriate ventilatory support is critical for symptom control, decreasing hospitalisations [11], and improving quality of life and survival in ALS. This has been clearly documented by a British randomised controlled trial: when ALS patients develop either orthopnoea, or a vital capacity <60% of predicted (or both), noninvasive ventilation (NIV) improves survival and quality of life [12]. These results seem to have had a major impact on referral of ALS patients for long-term mechanical ventilation (LTMV) by neurologists in the UK: an estimated 2.6%–3.5% of ALS patients were referred for NIV in 2000 [13], whereas close to 20% were found to be referred for LTMV in a similar survey performed in 2009, with a 72.5% success rate [14]. The vast majority of LTMV was noninvasive, with only 6.0% of all patients under LTMV having a tracheostomy (TPPV).

When discussing LTMV, the question of invasive ventilation (via a tracheostomy: TPPV), its timing, its relevance, its indication(s) remains problematic. In 1997, a Swiss multidisciplinary group focusing on the ethics of LTMV in neuromuscular diseases [15] stated that, for ALS patients, “invasive ventilation with the aim of prolonging life should not be recommended or started”. Reasons advocated for avoiding TPPV were: prolonging life in a highly dependent patient, major limitations in communication and a very heavy burden for caregivers.

In April 2010, a multidisciplinary approach for all cases of ALS in our area with regular follow-up consultations (every 3 months) was implemented. This article summarises the work of a multidisciplinary group on the question of invasive ventilation, 15 years after the recommendations of Kohler et al. [15], taking into account among other items published evidence, acquired experience and technical developments in LTMV.

Methodology

A workgroup on TPPV in ALS, including clinicians involved in the regular clinical activity of our ALS multidisciplinary consultation (Day-care centre, Division of Neurology, Geneva University Hospitals) and an ethicist from the Institute of Biomedical Ethics (Geneva University), was set up.

We first performed a systematic review of the medical literature using “PubMed” database, and seeking all publications in English and French using the MESH search items “amyotrophic lateral sclerosis”; “tracheostomy”; “non-invasive ventilation”, and limiting our search to clinical trials. We also analysed recent guidelines and recommendations on TPPV (American Academy of Neurology [AAN]), 2009; European Federation of Neurological Societies [EFNS] guidelines, 2012; Guidelines of the French Society of Neurology, 2006) [1, 16, 17].

We then invited representatives of homecare societies and a palliative care ward to discuss (1) the feasibility of TPPV in our area in terms of resources and financial support, and

(2) the difficulty of managing patients with advanced ALS and invasive ventilatory support.

Finally, we discussed practices related to TPPV with members of ALS centres in Switzerland (St-Gallen, ALS clinic; Nottwil, Centre for paraplegics; and Basel University Hospital), and two large ALS reference centres in France (Hôpital La Timone, Marseille, and Hôpital La Pitié-Salpêtrière, Paris).

This article summarises the results of our workgroup.

Published evidence (selected studies)

There is little doubt that NIV is effective in patients with ALS and orthopnoea and/or decreasing respiratory function, providing their bulbar function is normal or only moderately impaired [18]. Improved survival and Quality of life have been documented by a Cochrane review [19] and a randomised controlled trial [12], and NIV is recommended as a first-line treatment in patients with symptomatic respiratory dysfunction in the EFNS guidelines (2012) [16], the Practice Parameter Updates of the AAN (2009) [1] and the National Guidelines of the French Society of Neurology [17]. As far as TPPV is concerned, however, practices vary widely from country to country and from centre to centre: rates of TPPV in ALS range from 0% in a British series of 50 patients [20], 1.4%–14% in the US [1, 21], 3% in Germany [20], 2%–5% in France [22, 23], 10.6% in northern Italy [24] to 27%–45% in Japan [9].

Table 1 summarises some of the published findings regarding TPPV, rates of emergency implementation of TPPV, and rates of TPPV after advance care-planning in ALS. In a multicentre US survey, Moss et al. [25] studied 50 patients with ALS and LTMV. Forty-two percent had decided in advance to undergo LTMV while another 42% had LTMV initiated during an episode of acute respiratory failure (ARF). Twelve percent were considered “locked-in”; 40 (80%) were ventilated 24 hours/day. Of the 36 families who cared for a patient at home, 42% considered having the patient at home with LTMV as a “major burden, which changed their entire life and was stressful”. However, 83% of family caregivers would have encouraged their family member with ALS to choose LTMV again. On average, patients receiving LTMV at home were more satisfied than those living in an institution. The financial burden on families was high, but this is, at least to some extent, specific to the US system of healthcare.

Another US study from Ohio (Cazzolli et al. [26]) compared NIV (n = 25) with TPPV (n = 50) in patients with ALS. Only 8% of patients on TPPV had chosen TPPV in advance; all other tracheotomies were performed during emergency hospitalisations. Conversely, all patients on NIV had chosen electively their mode of mechanical ventilation (MV). Patients on TPPV and living at home were satisfied with their quality of life, whereas 28% of those living in an institution were unsatisfied. Family caregivers of patients with TPPV indicated that the burden of care was heavy. Conversely, management of secretions and relief of symptoms were problematic in NIV patients.

A group from Tennessee, US, compared patient acceptance and quality of life when on 24-hour TPPV between 8 patients with Duchenne’s muscular dystrophy (DMD) and 8

patients with ALS [27]. Patient satisfaction with TPPV was high in DMD, and significantly lower in ALS, suggesting that perception of TPPV varies according to the underlying disease and its prognosis.

Interestingly, in all of these relatively early studies performed in the U.S., TPPV was considered to be the “default” method of LTMV for ALS patients. More recent publications also suggest a low threshold for performing TPPV in certain European centres [28, 29].

A Japanese nationwide survey [30] included 709 ALS patients (2006), all under TPPV: 13% of patients developed a totally locked-in state (9% within 5 years after starting TPPV). Another Japanese study [31] describes 157 totally ventilator-dependent patients on TPPV, aged on average 61 years: 77% lived at home and 23% in a hospital. Family members were the only individuals to provide 24-hour support for 65% of these patients. Experiences with ALS patients in this country are unique because discontinuation of mechanical ventilation is illegal in Japan.

The largest study published, to our knowledge [24] (see table 1), was performed in northern Italy, and analysed prospectively all patients who had a tracheostomy performed among 1,260 ALS patients (n = 134; 10.6%) over a 10-year period (1995–2004). Tracheostomy was performed on average 2.4 years after diagnosis, electively in 56% and after an episode of ARF in 44%. Young male patients were more likely to have a tracheostomy than older or female patients, as were married subjects, which emphasises the importance of family caregivers (mainly the spouse). In-hospital mortality was high (20%), 65 (48%) returned home and 31% were institutionalised. Median survival was less than 1 year. Survival was related to age, follow-up by an ALS centre and marital status.

Two recent Spanish studies [28, 29] (see table 1) report a high rate of tracheostomy in ALS patients admitted to a respiratory care unit (RCU). A first prospective study included 136 ALS patients referred between 2004 and 2008: 28 patients had a tracheostomy (20%), 46% as an emergency procedure. The second study included all ALS patients re-

Table 1: Summary of selected studies of positive pressure ventilation via a tracheostomy in amyotrophic lateral sclerosis.

Study	Year, country	Type of study	No. of ALS patients included	Rate of TPPV (%)*	Emergency implementation of TPPV (%)	Proportion of patients who underwent TPPV after ACP	Other important observations
Moss et al. [25]	1996, USA	Cross-sectional, observational study	50	86%	42%	42%	12%: locked-in syndrome; 80% of patients on TPPV 24 hours/day
Cazzoli et al. [26]	1996, USA	Prospective observational study	75	67%	92%	8%	TPPV patients living at home (50%) satisfied with quality of life
Kaub-Wittemer et al. [33]	2003, Germany	Questionnaire survey of ALS patients under HMV	52	40%	Not applicable	19%	95% of patients on TPPV 24 hours/day
Hirano et al. [31]	2006, Japan	Questionnaire survey of ALS patients under HMV	157	100%	30.1	63.5%	77% of patients lived at home; 100% of patients on TPPV 24 hours/day; 6.4% totally locked-in; Mean time spent on TPPV: 4.4 years (SD 3.7 years)
Sancho et al. [29]	2010, Spain	Prospective observational study of patients admitted to a RCU	136	20.5%	46%	53.6%	45% of patients put on NPPV; 28% of patients on NPPV later switched to TPPV; 36% of patients on NPPV 24 hours/day; 64% of patients on TPPV used cuffless tubes
Chio et al. [24]	2010, Italy	Prospective observational study	1260	10.6%	44%	56%	Mean hospital stay after TPPV: 52 days; 20% in-hospital mortality after TPPV; 48.5% of patients discharged at home; 31% institutionalised after TPPV Mean survival after TPPV: 8.4 months
Vianello et al. [32]	2011, Italy	Retrospective observational study of unplanned TPPV	60	100%	100%	0%	70% of patients on TPPV 24 hours/day; 52% of patients discharged at home; Mean survival after TPPV: 30.3 months (SD: 4.4 months)
Sancho et al. [28]	2011, Spain	Prospective observational study of patients admitted to a RCU	116	33%	55%	45%	45% of patients on TPPV 24 hrs/day; Mean survival after TPPV: 10.4 months

* Among all patients studied; rate of TPPV is indicative and depends on study structure and inclusion criteria.

ACP = advance care-planning; ALS = amyotrophic lateral sclerosis; HMV = home mechanical ventilation; NPPV = noninvasive positive pressure ventilation; RCU = respiratory care unit; SD = standard deviation; TPPV = positive pressure ventilation via a tracheostomy

ferred to a RCU between 2001 and 2010 [28]: among 116 patients, a tracheostomy was indicated for 76 and 50% refused. Of the 38 tracheostomies performed, 55% were performed as an emergency procedure and 45% electively, mostly as a result of “failure” of NIV, “failure” of management of airway secretions or both. Noteworthy is the fact that prior to elective TPPV, 47% did not have NIV. Mean survival was 10.4 months, with a 1-year survival of 78.9%. No specific predictor of survival was identified.

Quality of life of patients and care-givers: burden of TPPV and prevalence of depression

In a retrospective chart review, Vianello et al. [32] (Padova, Italy) studied 60 ALS patients put on TPPV after an ARF (see table 1). For those evaluated, 15% were severely depressed and 85% estimated their quality of life as acceptable with a positive attitude towards TPPV.

A German questionnaire-based study described the health-related quality of life (HRQL) of 52 patients on LTMV and their caregivers [33]; 40% were on TPPV. Importantly, 81% of patients under TPPV had a tracheostomy without prior informed consent. HRQL scores did not differ significantly between NIV and TPPV groups: 94% of NIV patients and 81% of TPPV patients would have chosen their mode of ventilation again; 97% of NIV caregivers would advise NIV, but only 75% of TPPV caregivers would choose to do so. Interestingly, 30% of TPPV caregivers rated their own HRQL as lower than that of the patient.

Albert et al. [34] (New York, USA.) followed up 71 patients with advanced ALS until death or implementation of TPPV. The 14 patients who opted for TPPV were younger, had higher household incomes and used more supportive care. Caregivers of patients with TPPV reported greater depressive symptoms and a higher burden at baseline, but perception of burden and depression declined over time. In another study from Columbia University [35], at baseline 13% of caregivers had symptoms of major depression and 10% of minor depression (22% of ALS patients were depressed). The depression rate declined over time in caregivers to 10% (but remained stable in patients). The authors conclude that, although distress is not uncommon, clinical depression among caregivers of late stage ALS patients is the exception and does not increase as the patient’s death approaches.

TPPV in ALS: a few ethical concerns

Although physicians seem to base their decision to implement TPPV (or not) on an estimation of prospective quality of life [36, 37], they also tend to both under- and over-estimate their patients’ quality of life [36, 38]. The physician’s choice is therefore not an appropriate surrogate for the patient’s own assessment. Also, although the patient’s interests should legitimately be central, they are not alone in the picture, given the burden placed by TPPV on caregivers. Furthermore, the patients’ autonomy in their choices will be limited – here as elsewhere – by available health system resources. Finally, and most importantly, the rapidly progressive nature of ALS is likely to impact on the patients’ priorities and choices [38, 39]. The optimal timing of a self-determined decision thus becomes difficult to

determine. Advance care-planning is encouraged as early as possible, to provide time for reflection and integration of the choice within the patient’s priorities and life plans. In rapidly changing circumstances, however, decisions may become obsolete at a faster pace. This emphasises the importance of continuous shared decision making [40]. It also makes advance planning more challenging [41].

TPPV in ALS: comments from intensive care specialists

The intensive care specialist faced with the decision to admit to the intensive care unit (ICU) a patient with ALS may be confronted with two difficult decisions: that of performing a tracheostomy if the patient cannot be weaned from the ventilator (tracheostomies in ALS patients are often performed in an acute-care setting [24, 25, 28], table 1), and that of withdrawal of care if TPPV is refused by the patient. Despite the fact that, theoretically, the decision to implement TPPV should have been discussed beforehand, advance directives (ADs) as to cardiopulmonary resuscitation and TPPV are often not available when an ALS patient suffers from an episode of acute respiratory failure (ARF) (It is estimated that ADs exist for 30%–50% of cases of ALS with an ARF) [23]. If possible, admission to ICU, intubation and the possibility of long term TPPV with its consequences should be discussed with the patients and their family members together. Noteworthy is the fact that patients who are intubated in ICU can be awake and participate in such a discussion even if the environment is not optimal.

NIV is also increasingly used to improve weaning outcome and decrease the risk of reintubation after extubation in patients treated for an episode of ARF, although its efficacy in this setting is still a matter of debate [42–44]. To our knowledge, only one trial describes the successful use of NIV and aggressive mechanically assisted coughing for weaning off orotracheal intubation in neuromuscular patients after ARF (10% were ALS patients) [45]. Failure of NIV after extubation leads to either TPPV or palliative sedation. The fact that the option of TPPV exists does not necessarily mean that it should be implemented: ethics of intensive care have taught us that “not all that can be done should be done, but only what should be done” [46].

TPPV, ALS and palliative care

Medical interventions such as LTMV, while aiming to preserve the patient’s quality of life, also allow the disease to evolve to a more advanced stage with a progressive loss of independence and communication capacity.

The ability to communicate and to relate to others are key values in palliative care, but some patients may feel that life is worth living whatever happens and their preferences must be respected. Healthcare professionals must inform the patient of the options available, what benefit can be expected from their use, and also what risks and constraints each one entails. For TPPV some important issues are: the likelihood of being able to stay at home; the possibilities and modalities of external support; the burden of care for the family and the financial costs for the patient. Such conversations

can be distressing for the patient. As previously mentioned, they should be initiated early enough for the information to be imparted at the patients' own pace, to give them time to reflect and to leave them the opportunity to ask questions at a later occasion. Exploring and addressing the patients' fears is important. In particular, patients should be reassured as to the (very low) risk of choking to death, and palliative sedation should be explicitly discussed [47].

To make an informed choice about TPPV, patients also have to be aware of the risk of developing dementia and a locked-in state, which are associated with ALS, and severely limit decision-making capacity. In fact, the presence of cognitive impairment, sometimes very early in the disease, and the association between frontotemporal syndrome (up to half of patients with ALS) or frontotemporal dementia and ALS is increasingly recognised [1, 16, 48–50], and represents a challenge for the clinician when engaging in the process of advance care-planning. Frontotemporal dementia is associated with executive dysfunction, emotional blunting, loss of insight and behavioural changes [51]. The presence of cognitive dysfunction must therefore be systematically sought, to determine the patient's decision-making capacity. The absence of a consensus validated tools in ALS for the detection and assessment of frontotemporal dementia is a problem, and determination of a patient's decision-making capacity relies at least to some extent on the awareness and the experience of the healthcare professionals [1]. Family members can be of help for advance care-planning and when possible should be involved in the elicitation of the patient's goals and preferences, and in the discussion of therapeutic options.

The eventuality of interrupting TPPV should be discussed as well as the circumstances under which ventilation should be discontinued if the patient cannot decide for himself.

The experience and trends in three Swiss centres and in two large French ALS reference centres: illustrations of present trends

In Marseille's ALS referral centre, one of the 17 centres in France (Hôpital La Timone) approximately 350 ALS

patients, of whom 80 are under NIV, are followed up on a regular basis by a multidisciplinary team. Over the last 10 years, 53 patients have had a tracheostomy performed for LTMV, the majority after an episode of ARF (approx. 5% of all patients under LTMV). After tracheostomy, caregivers noted a transient global worsening in motor functions of their patients, partially explained by the lack of intensive physiotherapy. The trend in this institution is clearly to discourage TPPV, although all patients are informed of this possibility when informed about ventilator support; as such, elective TPPV has become a very rare procedure.

The Hôpital de la Pitié-Salpêtrière, ALS reference centre for the Ile de France region (Paris and surroundings), follows up approximately 1,200 patients. Out of these, 220 benefit from NIV and only a minority are treated with TPPV (21 patients). Here also, although patients are informed about the possibility of TPPV, almost all recent tracheostomies were performed after episodes of ARF.

Importantly, for both centres, as in all 17 ALS centres in France, the public healthcare system provides, if necessary, 24-hour support for TPPV patients who live at home, except for patients in whom disability linked to ALS appears after the age of retirement (60 years of age).

The three Swiss centres are: ALS Klinik, St Gallen; Nottwil centre for paraplegics; and Basel University Hospital, In St Gallen, over the past 10 years only a few patients were put on TPPV. The option of TPPV and its implications (especially caregiver burden) are openly discussed with patients and their caregivers (table 2). Most patients refuse TPPV because of the high caregiver burden. Trends are similar in Basel and in Nottwil (fewer than 5% of patients with TPPV).

Is there a profile for a "good candidate" for TPPV in ALS?

In a prospective study of 72 ALS patients, of whom 14 chose TPPV voluntarily, predictors of choosing TPPV were: younger age, having younger children, and higher income (the study was performed in the United States). Interestingly, religious beliefs were not predictive of choice [52]. The most important factors reported in the decision-

Table 2: Advantages and drawbacks of positive pressure ventilation via a tracheostomy (adapted from Andersen et al, European Federation of Neurological Societies 2012 guidelines [16], and Gonzalez et al. [23]).

<p>Advantages:</p> <ul style="list-style-type: none"> Increases survival Prevents aspiration of secretions of upper airway¹ Provides more effective ventilator pressures and better gas exchange² May improve symptoms of respiratory failure when noninvasive positive pressure ventilation fails Facilitates aspiration of secretions Avoids mask-related skin lesions and ulcerations "Frees" the face; allows use of glasses
<p>Drawbacks:</p> <ul style="list-style-type: none"> Generates more bronchial secretions Increases risk of infection Introduces risk of tracheo-oesophageal fistula, tracheal stenosis or tracheomalacia Greatly increases costs³ Increases family and carer burden, including 24-hour nursing requirement⁴ Ethical issues regarding discontinuation
<p>1: protection is partial, aspiration still possible since cuff is not always inflated; 2: adequate gas exchange is rarely a problem in noninvasive positive pressure ventilation and is not <i>per se</i> a main objective (as opposed to comfort); 3: highly dependent on medical coverage system; 4: difference probably minor when compared to totally ventilator dependant tetraparetic patients under enteral nutrition via percutaneous endoscopic gastrostomy.</p>

making process are: quality of life, severity of disability, ability to return home, ability to discontinue mechanical ventilation if they chose to do so, and concern about their family's quality of life [53]. Factors proposed to predict successful use of TPPV are [54]: a highly motivated patient who is empathic to the needs of the family, and engaged with the family and the community; a slowly progressive course of the disease; a thorough understanding of alternatives to TPPV, of the progression of ALS and the possible cognitive impairment; a well-informed family willing to assume the burdens of TPPV; financial resources to cope with the support network (this factor being healthcare system dependent); an advanced directive for discontinuing TPPV; and adaptability of both the patient and the family to constantly changing caregivers, equipment and limitations.

Managing TPPV in ALS at home in Switzerland: available resources

All subjects residing in Switzerland benefit from medical insurance coverage that will include expenses for the intervention of health professionals at home (10% is at the patient's expense). In urban areas at least, technical aspects of home ventilation and enteral nutrition are rather easy to organise and are managed at home by trained nurses. Finding trained specialists (otorhinolaryngologist, pulmonologist) willing to make home calls is often difficult and sometimes impossible. All nonqualified support for the patient (home cleaning, shopping for food, etc.) is at the patient's expense. Ensuring regular tracheal aspiration and maintaining airway patency relies on family caregivers, for whom no formal training is organised (and thus practices vary from centre to centre), which differs from what is offered in France, Germany or Scandinavian countries. Therefore the burden put on the family is considerable. Very few institutions are profiled for the intermediate or long-term care of these patients, and even elective short stays to offer a temporary relief to caregivers are difficult and challenging to organise. Patients can rely to some extent on financial support from patient associations or nonprofit organisations for neuromuscular diseases: this is, however, not automatic and discussed on an individual basis.

Discussion

On the basis of the medical literature, and practices in French and Swiss ALS expert groups, a certain number of observations can be made:

1. Although TPPV is feasible in ALS, with an acceptable quality of life, many studies suffer from a "centre bias". The "centre bias" refers to publications in which data suggest a low threshold for performing tracheostomy and, conversely, maybe a lesser experience with NIV. Indeed, TPPV was, until the end of the 1990s, considered in many centres as the "default" treatment for RF in ALS: most of these patients would probably be managed today on NIV. Using NIV 24-hours-a-day has become a usual practice in advanced ALS patients and is well tolerated [20, 29, 55–57]. Patients with ALS can be kept on full-time ventilation for at least 1 year, and often more with

appropriate management of interfaces and secretions. Strategies such as use of a mouthpiece during the day with a volumetric ventilator are clearly helpful [58, 59]. Increasing experience in NIV decreases the rate of NIV failure reported in previous studies, and substantially increases survival. Whenever a comparison is possible, and although quality of life under TPPV appears acceptable, quality of life scores under NIV are better than with TPPV.

2. Whenever TPPV is associated with being institutionalised, this has a clear negative impact on quality of life.
3. TPPV is presently most often initiated in emergency situations. Lack of prior informed consent and appropriate information is frequently reported. In fact, most patients with TPPV had an unplanned TPPV. Use of continuous NIV and mechanical cough assistance may in some cases allow weaning from orotracheal intubation and avoid TPPV [45].
4. There is a consensus on the very high burden that TPPV imposes on the caregivers, usually the family. "No caregivers are challenged more than those of ALS patients" [60]. Managing for months or years a patient with advanced ALS, under continuous NIV, with a gastrostomy and total dependence in activities of daily living represents a major burden on caregivers, and a high risk of burn-out must be considered if TPPV is suggested at this stage. Although a substantial part of home care for these patients is covered by medical insurance, nonmedical support is largely at the expense of the patient's family. When compared with neighbouring countries such as France and Germany, considerable progress has yet to be made in this field in Switzerland.
5. There is also a "geographical bias" in the decision to implement TPPV: the differences between Japan and the United Kingdom, for instance, are striking. This can result from cultural differences, differences in healthcare systems, laws or a combination of these factors. France, Norway, Sweden, and Germany have long experience with LTMV in neuromuscular patients, and the logistics and financial support available for home support are far greater than in many other European countries [61]. Exchanges with experienced Swiss and French ALS centres show that there is a clear tendency in our area to discourage TPPV in most cases, after openly discussing it with patients and their caregivers. This is corroborated by the low percentages of ALS patients put on TPPV.
6. The presence of cognitive impairment, which may sometimes occur very early in the disease, represents a challenge for the clinician when discussing advance care-planning, including the possibility of NIV or TPPV. Signs of neuropsychological dysfunction must be sought using appropriate tests on a regular basis.

Conclusions

TPPV is thus clearly a second line option, to be considered only when NIV fails and when prolonged survival is the goal and specific request of the patient (table 3). The grow-

ing awareness of cognitive disorders in the evolution of ALS is also a matter of concern. One exception is that of younger patients with predominantly bulbar disease, impaired clearance of secretions uncontrolled by medical treatment and failure of NIV: in these subjects, TPPV is a more clearly justified option [18].

However, reluctance to institute TPPV is strongly related to the absence of immediate hope in terms of therapeutic interventions and to the absence of communication or modes of empowerment of the patients on their environment when advanced in their disease. New technical devices for communication support such as Brain Computer Interfaces may change this in a rather near future, by facilitating communication and simple everyday tasks (reading, choosing music, videos etc.) [62]. This will strongly influence the beliefs and convictions of healthcare professionals. New therapeutic approaches may also change the prognosis and attitudes toward end-stage ALS, although unfortunately not in a near future. Last but not least, an integrated system of home care with specific training of providers, in order to alleviate the burden on caregivers, may change the focus of the discussion.

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Table 3: European Federation of Neurological Societies guidelines (2012) [16]: what do they tell us about positive pressure ventilation via a tracheostomy?

• NIPPV and, less frequently, TPPV are used to alleviate symptoms of respiratory insufficiency and prolong survival.
• NIPPV should be considered in preference to invasive MV (TPPV) in patients with signs or symptoms of respiratory insufficiency.
• TPPV has a major impact upon caregivers and should be initiated only after informed discussion [1].
• Unplanned (emergency) TPPV should be avoided through an early discussion of end-of-life issues, coordination with palliative care teams and appropriate advance directives [6].
• Ideally, the patient’s advance directives and a plan for management of respiratory insufficiency should be established before respiratory complications occur.

MV = mechanical ventilation; NIPPV = noninvasive positive pressure ventilation; TPPV = positive pressure ventilation via a tracheostomy

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