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RESEARCH ARTICLE

Tracheostomy ventilation in motor neurone disease: a snapshot of UK practice

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Abstract

Introduction: Motor neurone disease (MND) is characterized by rapidly progressive motor neurone degeneration which leads to muscle wasting. Mortality and morbidity are due to respiratory muscle failure which may be offset by ventilation. The aim of this observational study was to quantify the number and characteristics of patients living with MND choosing tracheostomy ventilation (TV) in the UK. *Methods*: Long-term ventilation services in the UK were invited to undertake a retrospective 5-year audit of MND patients under their care between April 2013 and March 2018 who had TV. Patient characteristics, the time spent on ventilation, hospital length of stay, discharge destination, and survival data were collected. *Results*: Sixty-eight MND patients were initiated on TV over the 5-year period. Eighty-one percent of patients received TV in an emergency setting with more than a third of these undiagnosed at presentation. Patients choosing elective TV were more likely to be male (85%) have a bulbar presentation (54%) and an increased survival of 10 months over the observation period. The mean length of hospital stay post TV was 136 days. Two-thirds of patients were discharged to their own home. *Conclusion*: Very few patients living with MND in the UK are currently receiving TV. In those who choose TV, there may be a survival advantage to planning an elective procedure. Despite the long inpatient stay and high care costs involved a majority of patients survived and were discharged to their own home.

Keywords: *Disease progression, motor neuron disease/therapy, respiratory insufficiency, tracheostomy, ventilator weaning*

Introduction

Motor neurone disease (MND) is a condition characterized by degeneration of motor neurones causing global muscle weakness. In turn, this leads to increasing disability, with possible dysarthria, dysphagia, and respiratory failure as muscles become denervated. Death is frequently related to respiratory muscle and cough failure (1). While neuroprotective therapies exist and increase life expectancy, noninvasive ventilation (NIV), if accepted can offset respiratory muscle weakness, reduce work of breathing, improve survival, and enhance quality of life (2–4). NIV is therefore recommended by the National Institute for Care Excellence (NICE) (5). Cough augmentation techniques are also recommended to clear secretions, maintain chest compliance, and prevent atelectasis.

While NIV may improve outcomes in MND, many patients, find the therapy onerous. Challenges surround the interface which may lead to pressure damage with prolonged usage and act as a barrier to communication. In addition, weakness of the upper airway muscles can cause laryngeal dysfunction, upper airway closure, and an inability to ventilate effectively by noninvasive means.

An alternative method of ventilating patients is with invasive ventilation via a tracheostomy. This requires the placement of an artificial airway anteriorly into the trachea between the cricoid cartilage and the suprasternal notch under sedation. Although there are risks associated with tracheostomy placement relating to both the procedure and the sedative medication, 80% of patients survive one year (6). Tracheostomy ventilation (TV) may

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also improve quality of life in MND patients (7–9).

TV is commonplace in some health care systems such as Japan, it is rarely used in United Kingdom where it is reported that approximately 1% of those with MND proceed to TV (10). Recently, a large longitudinal study of 190 MND patients receiving TV in Japan demonstrated that in experienced hands TV offered an increased survival of approximately 7 years compared to those who did not (4).

While there is some guidance on the use of TV in MND (11,12), NICE (5) does not comment on this. There is concern among health care professionals in the UK that TV patients have protracted inpatient stays, often waiting many months in hospital before being discharged into care homes. In addition, there is a perception that patients receiving TV may have a reduced quality of life and be at high risk of becoming "locked in" before death (13).

Although there is no mention of tracheostomy in the extensive NICE MND guidance, some UK centers do offer TV in MND while others do not. The objectives of this study were to determine both the number and type of patients with MND receiving TV, the circumstances in which tracheostomy is inserted, and the outcomes following tracheostomy insertion in the UK over a 5-year period.

Methods

This retrospective observational study was undertaken in July 2018 by the specialist home ventilation centers across the United Kingdom. Centers were approached via a number of networks (Home Mechanical Ventilation-UK, Specialist in Long term Ventilation at Home and British Thoracic Society) and invited to undertake a retrospective 5-year case note review of the use of NIV and TV in patients diagnosed with MND. Data were obtained by retrospective case-note review of patients set-up on TV between April 2013 and March 2018 inclusive. All patients receiving TV in MND were included; considering both elective and emergency tracheostomy insertion. Patients were included if they were subsequently found to have a diagnosis of MND after initial intubation or weaned from TV back onto NIV. Relevant permissions were obtained via the hospital audit and quality departments. Approval was granted via the Caldicott Guardian.

Data are presented as mean range for continuous, normally distributed variables, and % (n) for binary and categorical variables. Differences in length of stay, where available between elective and emergency tracheostomy were analyzed with the Mann–Whitney test. Kaplan–Meier's curves were used to assess long-term survival.

Results

Data were received on 68 MND patients set up on TV from 24 specialist UK HMV centers. Seventy-seven percent ($n = 52$) of these had a diagnosis of amyotrophic lateral sclerosis (ALS), 14% ($n = 10$) progressive bulbar palsy, and 9% ($n = 6$) were unknown or other. Fifty-one percent ($n = 35$) of all patients were NIV naïve before intubation. All elective patients received NIV before tracheostomy. Eighteen of the 24 centers initiated TV for patients with MND in the prior 5 years ending 31 March 2018, the remaining six centers had not. Twelve centers provided data on 2493 patients with MND. Sixty percent ($n = 1496$) of these opted for a trial of NIV. Eighty-three percent ($n = 1242$) successfully adapted to using NIV with 1.8% ($n = 22$) of these transferring to TV.

Tracheostomy ventilation

Patients receiving TV were more likely to be male (75% $n = 52$). Age at intubation was 59 years (range 32–82). Average time until intubation from diagnosis was 15.8 months (range 0–73). Those receiving TV electively did so later in the course of their disease than those who underwent an emergency procedure (24.9 months vs. 14.3 months). The majority of patients choosing TV were married, living with a partner or parent (71%, $n = 49$). This was similar in both the elective and emergency groups: (76%, $n = 9$) vs. (85% $n = 48$).

Emergency TV

Eighty-one percent ($n = 55$) of all tracheostomised patients were intubated as a result of an emergency admission. Only 36% ($n = 20$) of these had tried or been established on NIV before intubation. An acute admission for respiratory failure without a confirmed diagnosis of MND was the most common indication for long-term TV (36%, $n = 20$) in the emergency group. Not all emergency TV patients were managed with TV in the longer term. A wean was attempted in 54% ($n = 30$) of patients, a third ($n = 10$) of which were initially successful with eight patients being managed on NIV until death, two patients were reintubated and continued TV.

Elective TV

Nineteen percent ($n = 13$) of tracheostomised patients had opted for an elective procedure. In this group, bulbar dysfunction or upper airway difficulties were the main indications for having a tracheostomy (54%, $n = 7$). Only 18% ($n = 11$) of emergency procedures were undertaken for these

Table 1. Patient characteristics of those receiving emergency or elective tracheostomy ventilation.

	All patients	Emergency TV	Elective TV
Number of patients (*n*)	68	55	13
Gender male (%)	75.3%	69.0%	85.7%
Mean age at intubation (years)	59.0 (32–82)	59.0 (32–82)	58.5 (33–75)
Time from diagnosis until NIV (months)	16.9 (0–65)	18.8 (0–65)	15.0 (1–34)
NIV use pre-TV (%)	49 ($n=34$)	36 ($n=20$)	100 ($n=14$)
NIV usage pre-TV (months)	14.3 (0–59)	11.4 (0–59)	17.2 (4–51)
Time from diagnosis until intubation (months) (includes those diagnosed upon or post intubation)	15.8 (0–73)	14.3 (0–73)	24.9 (9–57)
Time from diagnosis until death (months) (if died before 31 March 2018)	26.6 (1–68)	23.76 (1–66)	37.6 (17–68)

NIV: noninvasive ventilation; TV: tracheostomy ventilation.
Results are reported as mean and full range.

indications. Males were more likely to choose elective TV (85%) (11 vs. 2) and had a tendency to be younger than their emergency TV counterparts (58.3 years vs. 61.3 years) (Table 1).

Hospital length of stay and place of discharge

For the 41 patients for whom data were available, hospital length of stay post initial intubation for all patients was a mean 136 days (range 40–564), with patients declared medically fit for discharge after 56 days (range 5–275). Elective patients had a shorter hospital length of stay (40 vs. 165 days, $p<0.001$). Similarly, for the 41 patients for which data were available, patients who elected for TV had a shorter time away from their intended place of care (61 vs. 178 days, $p<0.01$). Sixty-seven percent ($n=46$) of patients were managed in a critical care environment for all or most of their inpatient stay. Two patients died in hospital and a further two patients moved to a hospice for end of life care. The majority (60%, $n=38$) of patients returned home to their original residence, 27% ($n=16$) were discharged to a care home; seven of these on a temporary basis. Those who had a tracheostomy before a diagnosis of MND had a reduced chance of returning home with 53% returning to their original residence. Eighty-one percent of patients receiving TV were discharged home with a do not attempt resuscitation (DNAR) order; however, only 67% had advanced decisions to refuse treatment (ADRT) in place (Figure 1).

Once discharged home, patients were nearly all managed by a 1:1 tracheostomy trained skilled carer 24 hours per day. One patient had a 24 hour per day registered nurse package. The average number of carer hours per patient per day was 25.

Tracheostomy complications

In hospital. Twenty-two percent ($n=15$) of patients suffered complications when in hospital,

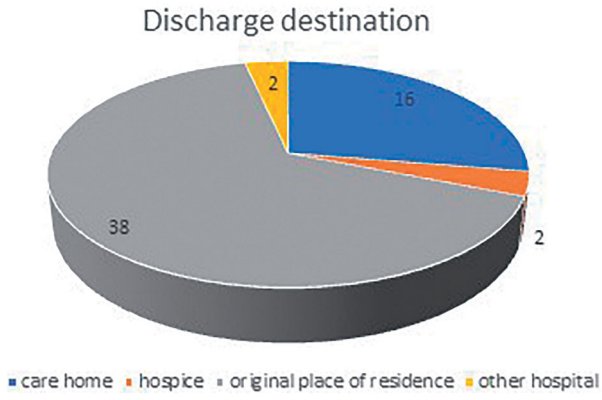

Figure 1. A summary of the discharge destinations from hospital care for patients receiving tracheostomy ventilation. Reported as number of patients (*n*).

half of these (53%) related to hospital acquired infection. One patient suffered cognitive impairment as a result of a hypoxic brain injury following an accidental ventilator disconnection.

Post hospital discharge. After discharge, 10 patients had complications relating to long-term ventilation via tracheostomy in the community, seven of these could be considered serious or catastrophic, five occurred in or on the way to a care facility. Three patients died of complications including a large granuloma causing airway obstruction, sputum plugging, and complications from a routine tube change. One patient became unconscious in the ambulance during a transfer home when a ventilator disconnection went unnoticed, they survived unharmed.

Post tracheostomy survival. By the end of March 2018, 25 patients (36%) had died. Twelve deaths were described as unexpected. One-year survival was 76% for all patients who received TV. There was no indication from the data, that any patients had died after becoming locked-in, but one patient

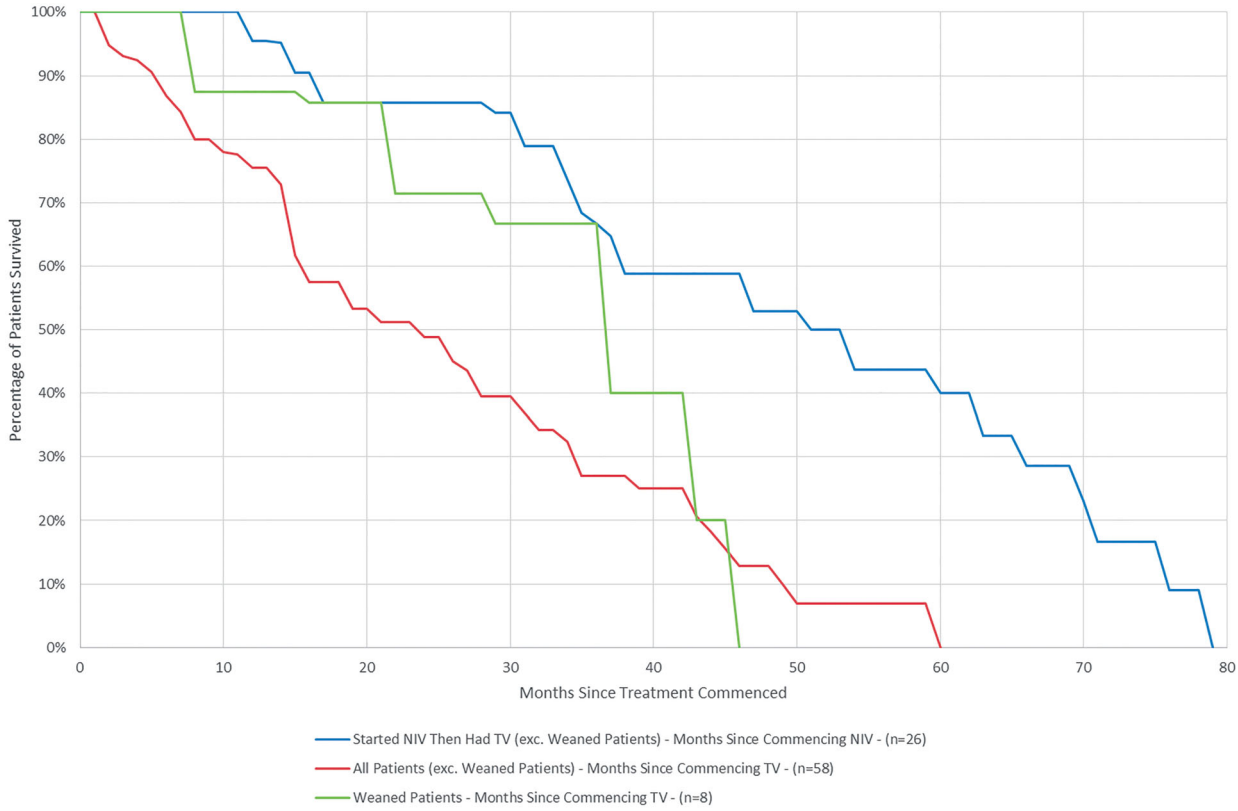

Figure 2. The median overall survival in three groups: those treated with TV who did not wean, those started on NIV who progressed to TV and those who received TV and were successfully weaned.

had ventilation withdrawn as per their ADRT when they become unresponsive.

For those patients who initially received NIV and then chose TV, 95% were still alive 1 year after initiating NIV. Median survival in this group was 53 months (Figure 2).

Median survival for patients who were not weaned and continued TV was 24 months (range 1–60) post intubation. For those eight patients extubated and successfully weaned onto NIV, the median survival post decannulation was 37 months (range 7–46). Only one of these patients had a diagnosis of progressive bulbar palsy, the remainder had ALS. For patients who chose elective tracheostomy, an increase in survival of 10 months was demonstrated compared to those receiving emergency TV with known MND (34 vs. 24 months). This was further improved when compared to those who received emergency tracheostomy before a formal diagnosis of MND (34 vs. 22 months).

Discussion

Main findings

Noninvasive ventilation. Within the limitations associated with a retrospective study, these data suggest that in keeping with NICE Guidance recommendations, (5) a majority of patients (60%) living with MND in the UK opt for a trial of NIV and that this is well tolerated in 83% of these patients. Furthermore, in this study, patients with MND survived 322 days on NIV prior to commencing TV. This would suggest that there has been an improvement in survival with the administration of NIV compared to that observed in Bourke et al.'s original study (2) where median survival was 216 days in patients receiving NIV. This may relate to increased awareness of the need for ventilator support in this patient group, earlier initiation of NIV, augmentation of NIV with insufflation–exsufflation devices, and/or improvements in the delivery of NIV over time.

Tracheostomy ventilation. The acceptance of TV is limited, with less than 1% of all patients living with MND receiving ventilation in this way. This is in marked contrast to some other economically well-developed countries where the incidence of TV in MND can be as high as 38% (10). The discrepancy may relate to differing health care systems and concerns actual or perceived in the UK surrounding delayed hospital discharge, the funding of ongoing care arrangements upon discharge in addition to the different ethical, moral, and legal arrangements in different countries.

There appears to be a male predominance toward electively choosing TV in accordance with the current literature (14). It has been suggested that this may be related to men being more accepting of invasive and life prolonging treatments than

women (13). Patients receiving TV were more likely to be in a stable relationship, which may also have contributed to the success of the therapy.

Elective tracheostomy. Elective tracheostomy insertion for long-term TV in MND patients is rare in the UK; this may relate to the lack of evidence around the survival benefits or improvements in quality of life. In keeping with the Spanish data (6), this study indicates a one-year survival of approximately 80%; with one quarter of patients surviving 3 years (15) post initiation of TV.

Sudden death accounted for almost 50% of deaths ($n = 12$). The high rate of sudden death is in keeping with the study performed by Sancho et al. (6) and may be explained by disturbances in autonomic and sympathetic activities (16). In this study, three sudden deaths were accounted for by complications or clinical incidents related to the tracheostomy, emphasizing the need for robust training of the carers who are delivering the complex care required by these patients in the community.

It is perhaps, not surprising that there appears to be a life prolonging effect of ventilating via tracheostomy in patients who have a disease in which death results from respiratory failure in around 80% of cases (1). If respiratory failure can be controlled adequately and the airway protected from aspiration then the patient may die of a non-respiratory cause, sudden, or otherwise. However, given that mean survival data in patients who were weaned from TV onto NIV appears to be enhanced when compared to those solely managed with TV it might not be the mode of ventilating but simply the adequacy of ventilation which prolongs life. These data suggest that there are differences between the phenotypes of MND with NIV potentially being preferable in patients with minimal bulbar and upper airway involvement who are able to protect their own airway. Eighty-eight percent ($n = 7$) of MND patients that successfully weaned from TV had ALS type of disease with only one patient having progressive bulbar palsy; this patient died 7 months post extubation. Furthermore, Japanese case reports by Tagami et al. demonstrated significant survival of many years in patients on NIV who were considering TV and declined (17). This further supports the proposal that it is the control of respiratory failure which prolongs life rather than the mode in which this is delivered.

Patients opting for elective TV appear to have improved survival over those who have emergency procedures, with mean survival in the elective group of 34 months rather than 24 months. There may of course be a selection bias in the type of patient wishing to consider TV who may be younger, have an improved performance status, be psychologically more robust with a "drive to survive" or receive more informal support from caregivers.

Over a third of emergency patients had a diagnosis of MND made after intubation. These patients may have been more unwell before invasive ventilation than those who opted for elective procedures with an increased mortality. NICE advocate that early diagnosis may enable earlier treatment and enhanced clinical outcomes (18). In addition, a lack of information and informed consent around living with TV in MND may lead to those having tracheostomy as part of an emergency admission being less prepared for living with TV. Withdrawal of ventilation maybe more likely than in those who have been able to make a considered decision (19). Further studies are recommended.

Discharge destination. Previous studies in the UK (20) and Italy (9) have demonstrated between 62% and 86% of those who received TV for respiratory insufficiency with MND were discharged to their own home. Fewer patients in this study 60% ($n = 38$) returned to their original place of residence. However, a further seven patients went into care homes on a temporary basis but the authors received no confirmation of whether these patients ever returned home.

Length of hospital stay, costings, and ongoing care needs. The mean length of hospital stay was more than a third of a year at 136 days with one patient spending more than 18 months in hospital awaiting discharge. The majority of the inpatient stay is within critical care, which can place a significant burden on a health care system already operating under significant operational and financial pressure. Fiscal concerns, of course do not end upon discharge. With health care provision for 1:1 care at home costing around £200k per year for a single patient (Premium Care Solutions, personal communication) it could be argued that managing TV patients with MND at home is not a cost-effective use of money. These monies could alternatively be used to provide many more patients with MND and/or other chronic diseases with services to enhance quality of life or offer improved survival. However irrespective of mode of ventilation, once MND patients progress to ventilator dependency, it is the reliance on ventilation (invasive or otherwise) for survival, the challenges this imposes on facilitating adequate communication and the patients' need for support in activities of daily living which necessitates the high cost of care.

Patients who opted for elective tracheostomy had a tendency to be discharged earlier (40 vs. 165 days), this may be related to a more planned approach facilitating prior funding for care packages, appointing, and training carers prior to admission. These data would lend support to

tracheostomy insertion occurring before patients become unwell and require emergency procedures to manage acute respiratory failure. There are however significant regional variations in the acceptance of this method with many clinical commissioning groups declining to approve preemptive funding.

Only 67% of those discharged had an ADRT in place. TV does not slow the progression of MND and patients can lose the ability to communicate. Advanced care planning before discharge including ADRT and appointing a lasting power of attorney is an essential part of decision making to ensure patients' wishes are respected in the future.

Study limitations

This is a retrospective study and thus has associated biases including missing data and increased data input errors that were collated by different persons from multiple study centers. The numbers of patients involved in some aspects of this study are small for example with only eight patients being successfully weaned from invasive mechanical ventilation and only 13 patients receiving elective tracheostomy. This is however a true reflection of the current status for patients living with MND in the UK. This study does not review the important outcomes of quality of life measures for patients requiring or wanting TV, nor for their informal carers, who often rate their own overall quality of life as lower than that of the patient (7).

Conclusion

This is the first study to survey the provision of long-term home mechanical ventilation use including tracheostomy provision for patients living with MND in the United Kingdom. These data support improved survival in patients receiving NIV in comparison with data from 2006. These data also suggest favorable survival outcomes for patients receiving TV and imply that patients can be discharged back home from hospital with appropriate packages of care. The costs of this however are significant. Patient hospital length of stay in a critical care environment is frequently prolonged but if managed effectively and proactively can be reduced to weeks.

Overall, this study emphasizes the need for research with prospective quantitative and in-depth qualitative studies to help shape future iterations of the NICE MND Guidance. Such guidance is urgently required to help guide professionals when confronted by patients who asked for TV to manage the respiratory complications of MND and to support patients who wish to make informed choices about their future care.

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Declaration of interest

There are no competing interests for any author.

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