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Delivery of noninvasive ventilation to people living with motor neuron disease in the UK

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Shareable abstract (@ERSpublications)
There is significant variation amongst UK healthcare professionals and services in the way noninvasive ventilation is delivered to people with motor neuron disease. Addressing weaknesses in all aspects of respiratory care could lead to improved outcomes. https://bit.ly/3V7Ru1x

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Abstract

Objective Noninvasive ventilation (NIV) improves survival and quality of life in motor neuron disease (MND), but many patients fail to receive effective ventilation. This study aimed to map the respiratory clinical care for MND patients at a service and individual healthcare professional (HCP) level to understand where attention may be needed to ensure all patients receive optimal care.

Methods Two online surveys of HCPs working with MND patients in the UK were conducted. Survey 1 targeted HCPs providing specialist MND care. Survey 2 targeted HCPs working in respiratory/ventilation services and community teams. Data were analysed using descriptive and inferential statistics.

Results Responses from 55 HCPs providing specialist MND care who worked at 21 MND care centres and networks and 13 Scotland Health Boards were analysed from Survey 1. Responses from 85 HCPs from respiratory/ventilation services and 73 HCPs from community teams, representing 97 services, were analysed from Survey 2. Significant differences in practice were identified at each stage of the respiratory care pathway as well as evidence of the need for improvement. This included when patients were referred to respiratory services, the time taken waiting to commence NIV, the availability of sufficient NIV equipment and provision of services, particularly out of hours.

Conclusion We have highlighted significant disparity in MND respiratory care practices. Increased awareness of the factors that influence NIV success and the performance of individuals and services is important for optimal practice.

Introduction

Motor neuron disease (MND) is a progressive neurological condition. Death usually occurs within 2–3 years of symptom onset, and the most common cause of death is respiratory failure [1]. The only intervention that substantially improves survival and sustains quality of life is noninvasive ventilation (NIV) [2]. However, many patients have low adherence, and even where adherence is good, ventilation is not effective in correcting hypoxia in many patients, which is leading to poorer survival rates [3, 4]. The success of NIV depends on many factors throughout the respiratory care pathway; from diagnosis and preparing for NIV, initiation, monitoring and optimisation, and end-of-life care [4, 5]. Current guidelines provide recommendations, but they are fragmented and do not adequately cover all aspects of the pathway [6–12].

There is a need to better understand how NIV is delivered to people living with MND (plwMND). This study aimed to describe the current practices of individual healthcare professionals (HCPs) and services providing specialist MND care, respiratory/ventilation services and community services that support NIV delivery in the UK.

Material and methods

A multi-method design [13] was used involving two online cross-sectional surveys, sequentially, using Google Forms (https://docs.google.com/forms) and Qualtrics (www.qualtrics.com) (see supplementary material). The questions for the surveys were informed by our earlier research [4, 5] and existing clinical guidelines (in particular, the UK National Institute for Health and Care Excellence (NICE) 2019 guidelines) [6–12], as we wanted to explore whether current recommendations are being met. Survey 2 was also informed by the findings of Survey 1. The data were analysed using statistics with SPSS Statistics for Windows Version 25.0 (IBM, SPSS Inc., Armonk, NY, USA). Comparisons (*e.g.*, between size of services) were performed using the Fisher's exact test. All comparisons were two-tailed, and a p-value of $\leqslant 0.05$ indicated significance.

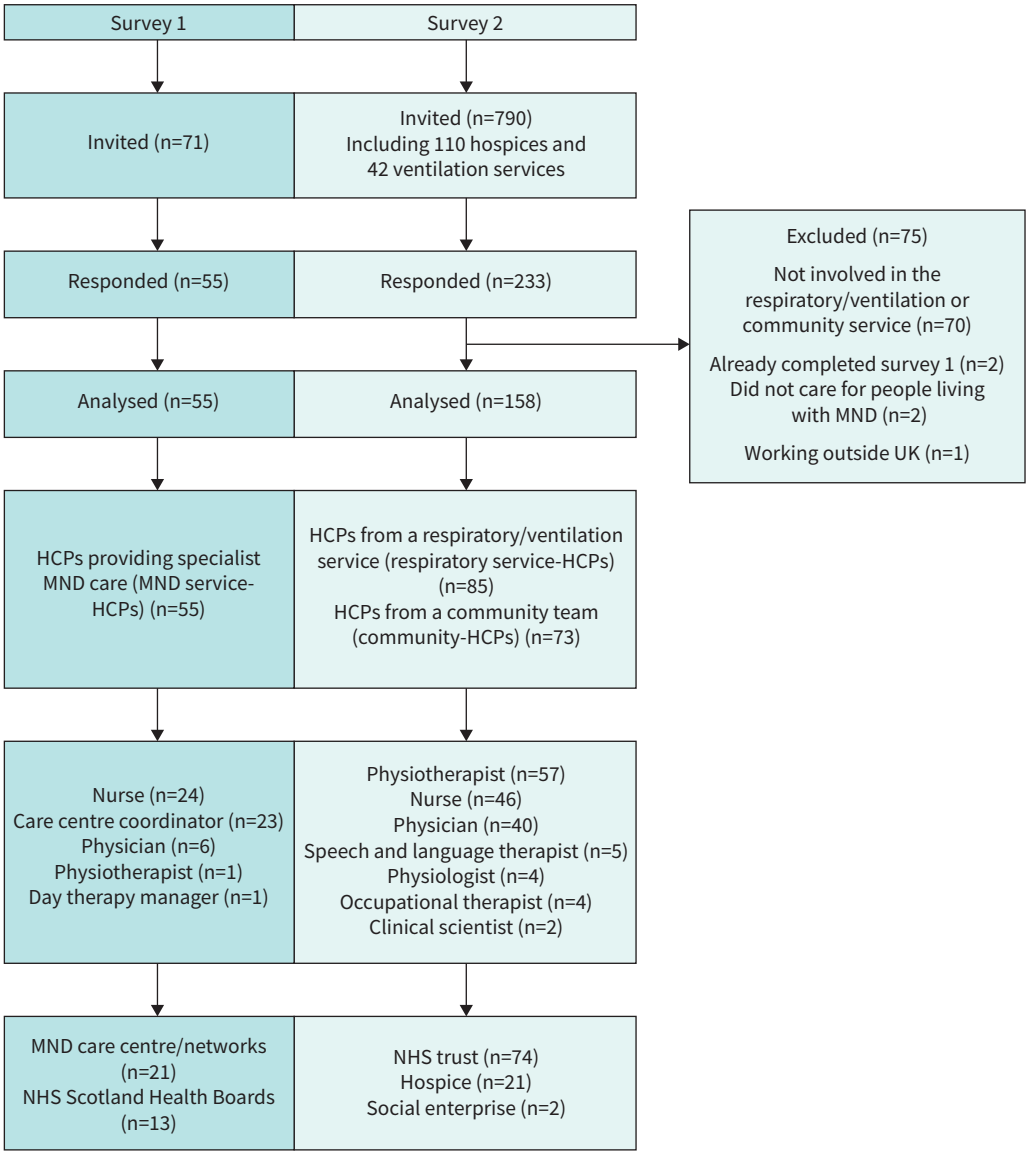

FIGURE 1 Flowchart of data collection and participant characteristics. HCPs: healthcare professionals; MND: motor neuron disease; NHS: National Health Service.

Survey 1

Survey 1 targeted HCPs providing specialist MND care in the UK (referred to as MND service-HCPs). A purposive sample involving MND care centre coordinators as gatekeepers and snowballing techniques was used to recruit participants. This involved existing participants sharing the survey with other people. People were invited to complete the survey *via* e-mail/telephone, and the survey was open for 1 month in April 2018. Ethical approval was obtained from the University of Sheffield (Ref. 018519).

Survey 2

Survey 2 targeted two groups: HCPs working in respiratory/ventilation services (staff who identified themselves as being involved in decision-making about the technical aspects of delivering NIV), referred to as respiratory service-HCPs, and HCPs working in community teams, referred to as community-HCPs. Two clinical vignettes and a core set of questions applicable to all participants were used. A subset of questions relating to technical aspects of NIV delivery were given to respiratory service-HCPs.

A convenience sampling approach was used by inviting staff working in services identified in Survey 1 as delivering NIV care, staff identified on hospital websites, and through personal clinical contacts. The survey was also advertised on social media, through charity networks/newsletters and using snowballing techniques and was open for 9 weeks from January 2019. Approval was obtained from the Health Research Authority, UK (IRAS ID 254661).

Results

Figure 1 provides a flowchart of the surveys and the characteristics of participants. The 55 MND service-HCPs in Survey 1 had an estimated total caseload of 4547 patients. The 158 Survey 2 participants worked for 97 services which included 47 of the services (92%) identified in Survey 1 as providing NIV. 54 services were specialist respiratory services. Responses describing services were excluded if there was variation between individuals replying from the same service. Services were categorised according to the number of plwMND using NIV as small (20), medium (20–50) and large (>50). Further data are included in the supplementary material.

Timing of involvement of respiratory/ventilation services (both surveys)

Over half of HCPs working in MND services (n=30; 55%) reported that referrals to respiratory/ventilation services occurred at the onset of respiratory signs/symptoms. HCPs working in the respiratory services reported that the most common time point of referral was when the patient developed signs, symptoms and/or respiratory function decline (n=33; 40%). Five respiratory service-HCPs (6%) reported that referrals most commonly occurred at the onset of respiratory failure. According to respiratory service-HCPs, 21 services (total number of services 54, 39%) received referrals at the time of MND diagnosis. 24 respiratory service-HCPs (29%) thought that referral occurred late, and of those who said the most common time point of referral was at diagnosis, nine (75%) thought this was at the right time. In 28 MND services (51%), respiratory specialists were available to see patients prior to the development of respiratory signs/symptoms. However, respiratory service-HCPs most commonly worked in a service that was separate to the MND clinic (n=52; 63%).

Respiratory function monitoring (Survey 2)

The majority of services (n=50; 98%) used more than one respiratory test with 41 services (80%) using five tests or more. One service (2%) reported using 12 different tests. The most commonly used respiratory test was forced vital capacity (FVC) (41 services; 77%) (figure 2). Polysomnography was used by 15 services (28%). Indications for polysomnography included if the patient was experiencing sleep apnoea, sleep disturbance or bulbar dysfunction and when other tests were inconclusive. Despite the NICE guidelines recommending the use of nocturnal oximetry and/or a limited sleep study in uncertain cases, eight services (15%) used neither of these tests [6].

Respiratory service-HCPs indicated the respiratory function threshold values which helped them decide whether to recommend NIV (table 1). Staff tended to report similar thresholds whether patients had respiratory symptoms or not. For those without symptoms staff were using a higher FVC (median 70% predicted) than that stated in the NICE guidelines (<50% predicted) [6]. There was variability in individual answers and when collated at service level. Respiratory service-HCPs commented that testing could be inaccurate, and that (in line with clinical guidelines) decision-making relied on a global assessment of tests, symptoms and patient choice. In those with bulbar dysfunction, HCPs reported placing more significance on symptoms, polysomnography and other measures of ventilation (*e.g.*, blood gases).

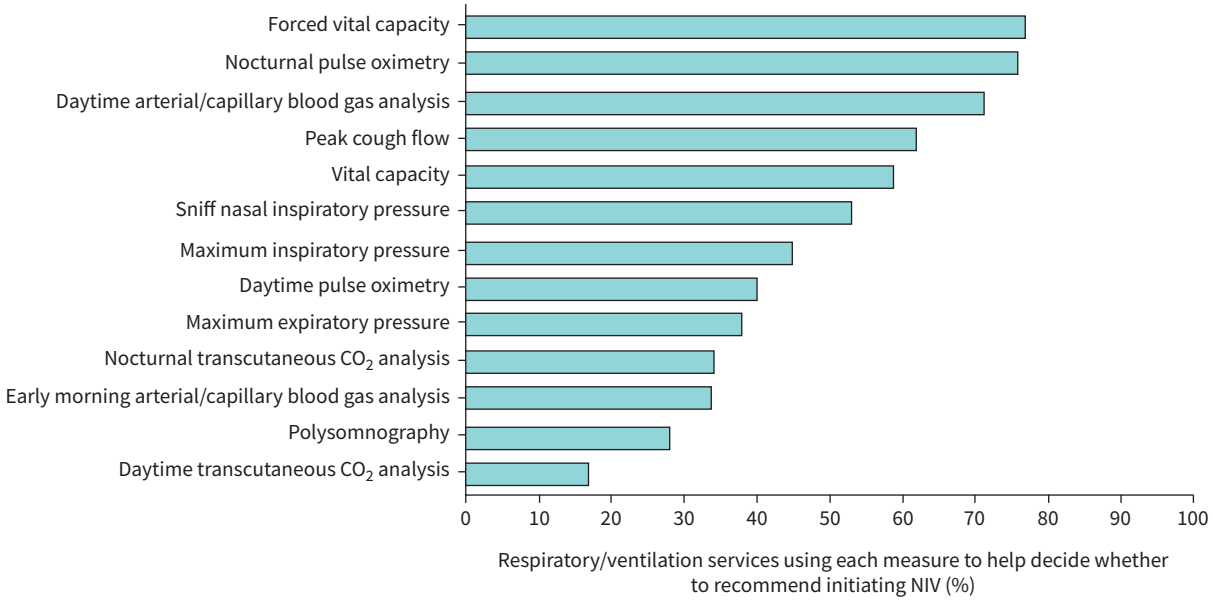

FIGURE 2 Respiratory function tests used by respiratory/ventilation services (n=53). NIV: noninvasive ventilation.

Discussions about NIV (both surveys)

The vast majority of Survey 2 participants (n=151; 96%) had a role in discussing the potential need for NIV. Discussions often start after diagnosis but before the onset of respiratory signs/symptoms or tests (n=73 out of 116; 63%). 43 (38%) stated this was the most common time point in their experience (figure 3). When discussions occurred most commonly before the onset of respiratory signs/symptoms, 33 HCPs thought this was the right time (n=33 out of 43, 77%). However, when discussions occurred most commonly at the onset of respiratory failure, most HCPs (n=6 out of 7; 86%) thought this was too late.

Respiratory service-HCPs stated who would provide information to patients about NIV (figure 4). The respiratory/ventilation services covered most topics but particularly technical aspects of using NIV. More than half of the MND services covered some aspects of the benefits prior to referral. However, respiratory service-HCPs working at 36 services (69%) reported that the MND team did not talk about the impact of NIV on carers, and respiratory service-HCPs working at 37 services (71%) reported that the MND team did not talk about options around withdrawal/end-of-life. Respiratory service-HCPs in 13 services (25%) reported that options around withdrawal/end-of-life were not discussed in their service either. Respiratory service-HCPs in four services (8%) said patients are often given limited (or no) information by the MND team.

Timing of initiation of NIV

NICE guidelines recommend patients with daytime hypercapnoea be seen within 1 week of referral [6]. For urgent referrals, 31 respiratory service-HCPs (39%) reported that patients usually have to wait <1 week to

TABLE 1 Thresholds used by services to decide whether to recommend a trial of noninvasive ventilation for patients without bulbar dysfunction

Respiratory function test	With symptoms of respiratory insufficiency, median (range) mode (NICE value)	Without symptoms of respiratory insufficiency, median (range) mode (NICE value)
Forced vital capacity %	70 (50–100) 80[#] (80)	70 (40–80) 50[¶] (50)
Vital capacity %	50 (40–80) 50 (80)	50 (40–80) 50 (50)
Sniff nasal inspiratory pressure cmH_2O	50 (4–65) 65[#] (40)	40 (4–65) 40[¶] (65 men/55 women)
Maximum inspiratory pressure cmH_2O	60 (30–80) 50 (40)	40 (20–80) 40 (65 men/55 women)
Maximum expiratory pressure cmH_2O	60 (30–113) 60	60 (30–113) 30

The recommended thresholds from the National Institute for Health and Care Excellence (NICE) guidelines are also included for reference [6]. [#]: two services excluded from the analysis due to variation in responses given by two respondents working at each service; [¶]: one service excluded from the analysis due to variation in responses given by two respondents working in the same service.

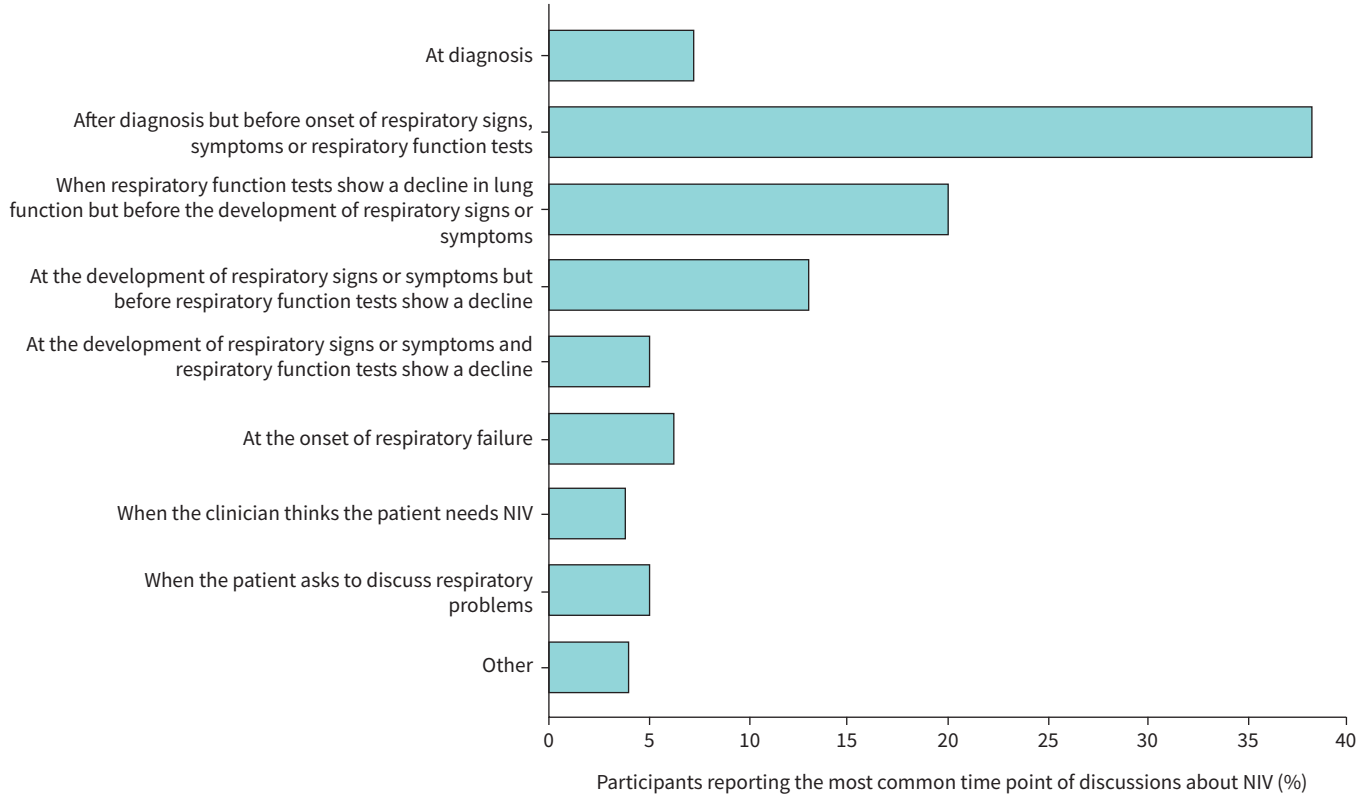

FIGURE 3 Most common time points of discussions about noninvasive ventilation reported by Survey 2 participants (n=112). NIV: noninvasive ventilation.

see the respiratory team. 22 (28%) said the wait was usually 1 week, 22 (28%) said the wait was usually 2 weeks, one (1%) said the wait was 3 weeks and three (4%) said 4 weeks. There was no significant association between size of the service and appointment waiting time for an urgent referral (Fisher's exact test, p=0.677). For non-urgent referrals, 4 weeks was the modal time (32%) but 18 (24%) reported waits of 5 weeks or more with four (5%) reporting waits of 12 weeks. There was no significant association between size of the service and appointment waiting time for a routine referral (Fisher's exact test, p=0.250). 32 services (78%) had a waiting time of <1 week to commence NIV. 10 services were excluded from this analysis due to variation in responses from individuals working at the same service (two per service). Five services (12%) had a waiting time of 2 weeks, two services (5%) had a waiting time of 1 week and two (5%) had a waiting time of 4 weeks. There was no significant association between size of the service and waiting time to commence NIV (Fisher's exact test, p=0.212).

Locations of initiating NIV (Survey 2)

The most common location available and used for initiating NIV was an outpatient setting with 38 services (79%) using this location. Initiation as a multiple-night admission was used in 31 services (65%), as a one-night admission in 19 services (40%) and as an inpatient day-case in 17 services (35%). Domiciliary initiation was offered in 31 services (65%). HCPs preferred initiating patients as an outpatient (n=33; 46%) followed by patient's homes (n=15; 21%). Nine services (19%) reported no funding for domiciliary initiation.

Equipment provision and funding (Survey 2)

Figure 5 shows the amount of equipment provided by services. In total, only 20 services were able to provide at least two NIV machines, one battery pack, two masks per year and a humidifier. Staff working in four services (15%) reported that no NIV machines were funded, with one HCP reporting charity funding was required. All four services had <20 plwMND using NIV in their service. Eight services (32%) had no funding for battery packs. Only one of these services had >20 plwMND using NIV in their service. Three services (12%) had no funding for masks and four (15%) had no funding for humidifiers. All of these services had <20 plwMND using NIV in their service.

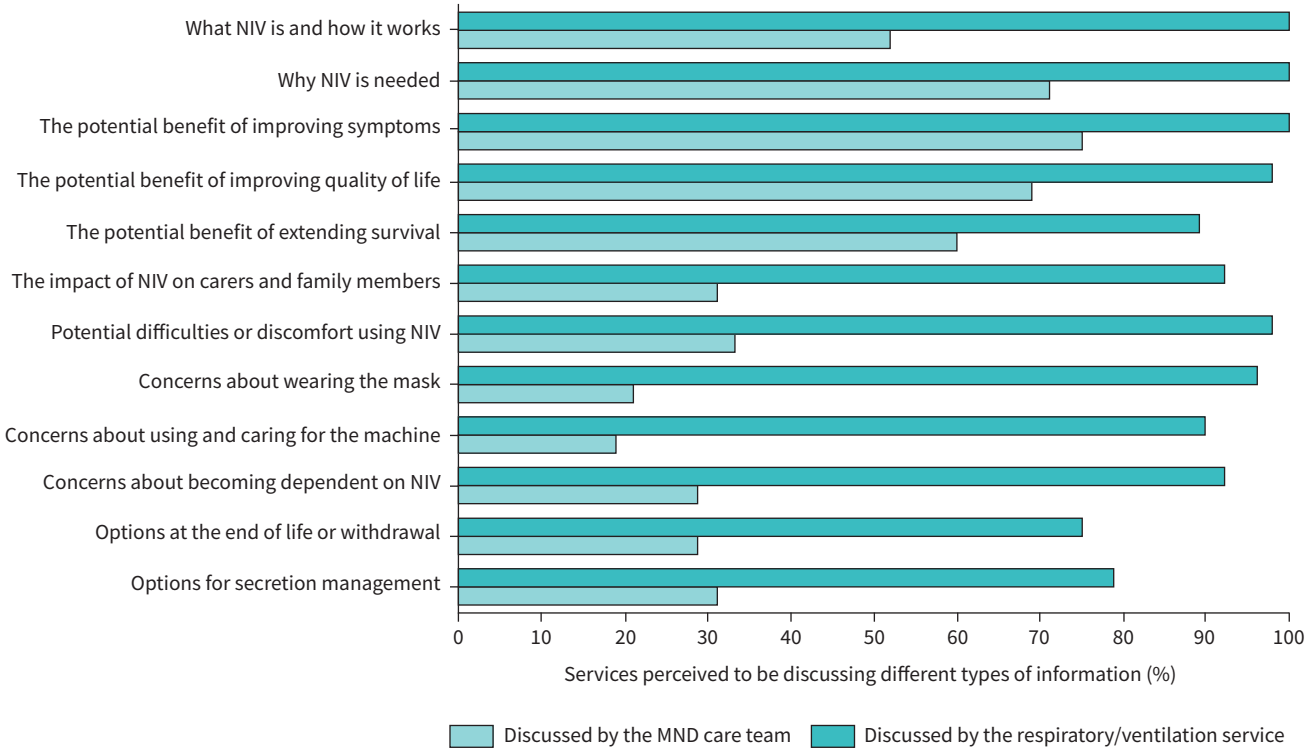

FIGURE 4 Information perceived to be given by the motor neuron disease care team and respiratory/ventilation service to patients from the perspective of respiratory service-HCPs (n=52). NIV: noninvasive ventilation; MND: motor neuron disease.

Pressure-targeted settings were used by 44 services (96%). Volume-assured pressure support was used by 23 services (50%) with inspiratory positive airway pressures of 12 cmH_2O and expiratory positive airway pressures (EPAP) of 4 cmH_2O being the most common choices for initiating NIV. The most common preferred choice for initiating both a patient with and without bulbar symptoms was pressure-targeted (n=54 (76%) and n=58 (82%) respectively).

A spontaneous-timed mode was available to use in 42 services (89%). 10 services (21%) used a spontaneous ventilation mode, 10 (21%) used a timed mode and 11 (23%) used other modes such as intermittent positive-pressure ventilation. The most common preferred ventilation mode was spontaneous-timed (n=51; 76%).

Services had a variety of mask interfaces available, including nasal pillows (n=48; 100%), total face masks (n=47; 98%), nasal masks (n=47; 98%), oronasal masks (n=46; 96%) and mouthpieces (n=34; 71%). For patients with and without bulbar symptoms, the most common choice of mask for initiation was an oronasal mask (both n=31; 44%) followed by a nasal mask (n=26 (37%) and n=21 (30%) respectively). Mask selection depended on patient choice, disability and mask fit.

Early initiation and follow-up (both surveys)

The daily target for using NIV recommended by respiratory service-HCPs ranged from 4 to 12 h with the most common being 4 hours (n=14; 35%). The most common preferred target representing optimal adherence for patients with and without bulbar impairment was using NIV all night (n=39 (51%) and n=44 (57%) respectively), although HCPs often wrote that targets were individualised. 16 respiratory service-HCPs (20%) stated that they recommend patients try to increase their usage of NIV. Strategies to achieve this included encouraging patients to use NIV during the day to begin with before moving to night-time use.

Following initiation, the median number of weeks to the first respiratory follow-up was 2.5 weeks (IQR 1–4), to the second follow-up was 8 weeks (IQR 4–12) and to the third follow-up was 12 weeks (IQR 6–24). The median time for routine follow-ups after the patient is established on NIV was every 12 weeks (IQR 12–12). Community-HCPs were asked to state how often they saw patients using NIV, which ranged from 1 to 24 weeks.

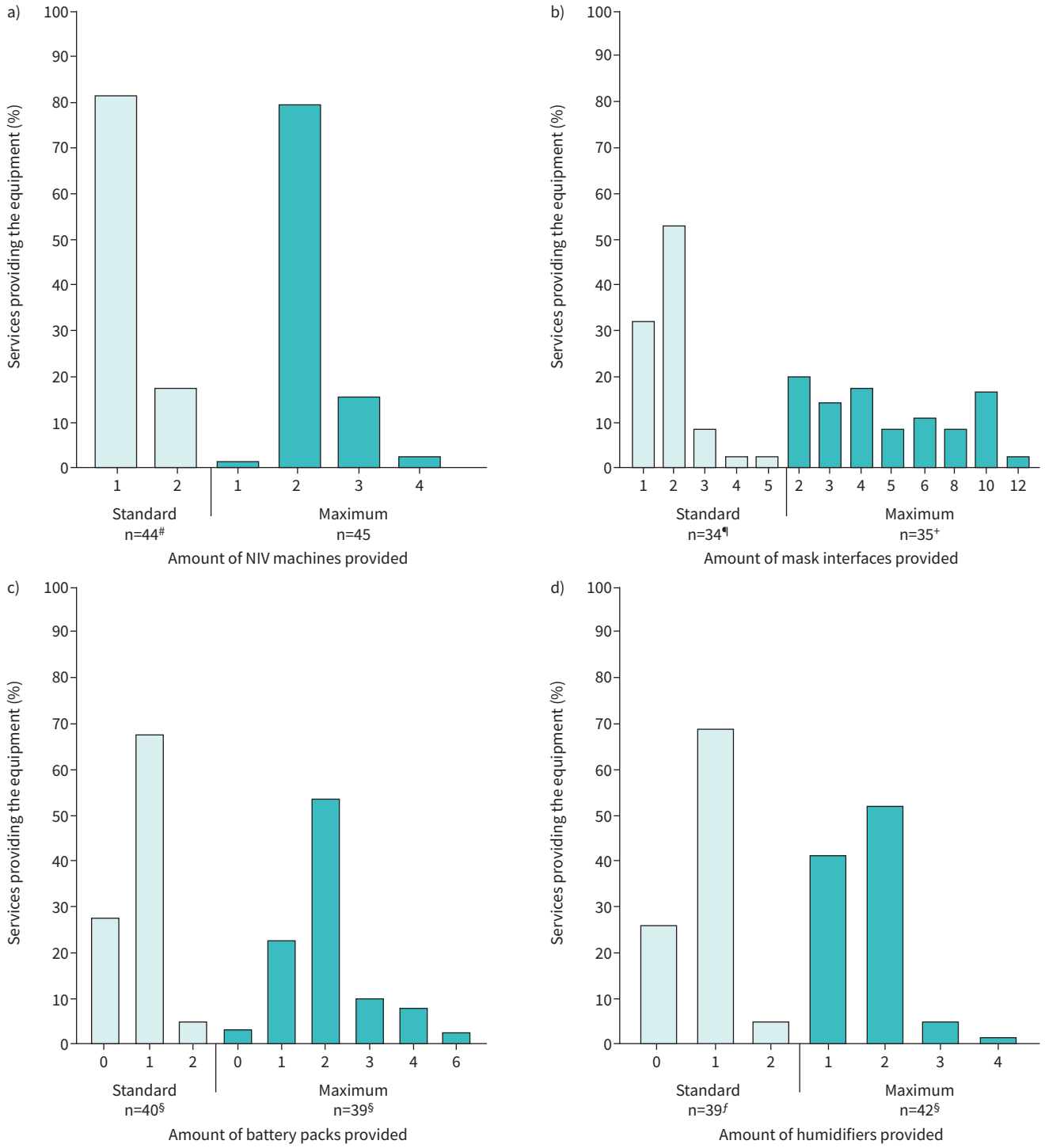

FIGURE 5 Amount of equipment provided by respiratory/ventilation services: a) NIV machines, b) mask interfaces, c) battery packs and d) humidifiers. NIV: noninvasive ventilation. #: three services excluded from the analysis due to variation in responses given by two respondents working at each service. For example, one participant stated that the standard amount of NIV machines provided in their service was one and another participant working in the same service stated that their service provided two NIV machines as standard; ¶: 13 services excluded from the analysis due to variation in responses given by respondents working in the same service; +: nine services excluded from the analysis due to variation in responses given by two respondents working at each service; §: four services excluded from the analysis due to variation in responses given by two respondents working at each service; f: five services excluded from the analysis due to variation in responses given by two respondents working at each service.

Once established on NIV, most MND service-HCPs monitored the patient for symptoms of respiratory insufficiency (n=52; 95%) and NIV comfort (n=47; 85%). Respiratory services represented in Survey 2 reported ways in which they monitored effective adherence and ventilation. 16 (21%) services used patient-reporting alone to monitor adherence, but others used machine downloads (n=51; 66%) and/or telemonitoring (n=25; 33%). 13 services (18%) used patient-reported symptoms alone to monitor ventilation. Others used a combination of symptoms and objective measures: oxygen/CO_2 measurements (n=49; 69%), machine downloads (n=44; 62%), telemonitoring (n=23; 32%) and polysomnography (n=9; 13%).

In-hours telephone support was available in 81 services (84%) and e-mail support was available in 55 services (57%). 63% of services (n=57) could be contacted by patients during out-of-hours times (outside of Monday–Friday 9:00–17:00). Funding was a barrier to out-of-hours support, but staff also identified that those answering calls out of hours were not always adequately trained to address problems.

Modifications to NIV therapy to improve adherence/effectiveness of NIV (Survey 2)

Participants reported that the pressure levels used were dependent on weight, comfort, tolerance, efficacy and bulbar function. Individual patient adaptations included higher EPAP and longer breath length for patients with bulbar dysfunction. A shorter rise time was reported to reduce airway collapse.

Figure 6 shows what troubleshooting steps services used to overcome mask leaks. The most common step taken was optimising mask fitting (n=58; 94%). To overcome upper airway obstructive events, the most common step taken was increasing the EPAP, which was used by 46 services (77%). Ventilator setting changes were triggered by patient discomfort, poor adherence/compliance, inadequate ventilation/asynchronies and respiratory decline.

End-of-life care (Survey 2)

Survey 2 participants reported the most common time of discussion about end-of-life respiratory care is when the patient asks to discuss end-of-life care (n=97; 69%) and when there is increased dependency on the ventilator (n=91; 65%). Only eight HCPs (6%) reported the most common time being when a patient is initiated on NIV. 50 participants (37%) thought discussions occurred late. This was particularly the case for those who said discussions occur most commonly when there is increased dependency on the ventilator (n=16; 70%). Participants commented that discussions should be patient-centred and occur early as part of a process to allow patients the time to plan. One person thought planning end-of-life care too early could impact on the success of NIV.

Services used a variety of supportive measures when discussions about the withdrawal of NIV began including discussing palliative care (n=67; 97%) and offering reassurance (n=65; 94%). 69 participants (72%) thought patients were referred to palliative care services at the right time, but 20 (21%) thought referral was late.

Discussion

Despite NIV being the most effective treatment for MND [2], our research demonstrates variation in clinical practice in the UK. We have identified sites and individuals who have the skills, equipment and

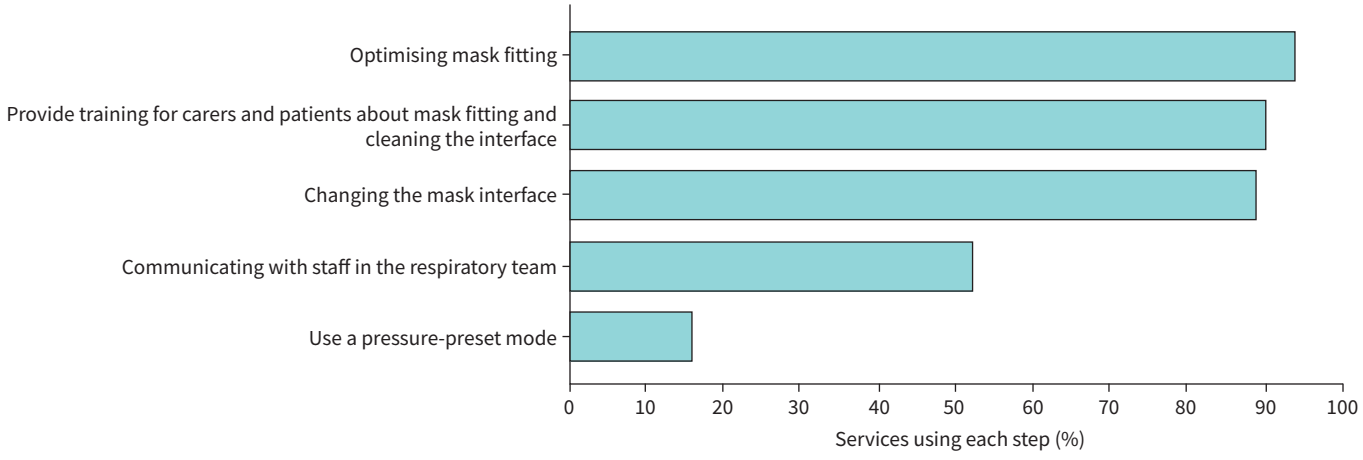

FIGURE 6 Steps taken by services to overcome mask leaks (n=62).

staff to deliver best practice in at least some areas of the respiratory pathway. However, this is not yet available to all patients at all sites. Variation occurs due to individual preference and service limitations, and specific evidence-based quality standards to guide practice remain limited. Published guidance [6, 8, 9] particularly lacks detail about the technical aspects of how NIV is initiated and optimised and how patients should be followed up. This might explain why this aspect of services was so variable. We have made some recommendations based on the best available current evidence [4–12], along with evidence of good local practice and our consultations with experts, that could be considered to help services evaluate and improve their care.

While the evidence for optimal timing of NIV is limited [14], early focus on respiratory function enables time for patients and carers to be optimised physically and practically [15]. Patients need to understand the trajectory of their disease and their options in order to make the "right decision" at the "right time" [16]. Moreover, there may be a survival benefit for early NIV initiation [17]. Our research found evidence of good practice with many services preparing patients for respiratory failure shortly after diagnosis [6]. However, as reported elsewhere [18, 19], HCPs in our study reported that discussions about respiratory function and referrals to respiratory/ventilation services were often taking place at the onset of respiratory failure. It was reported in some services that patients were often given limited (or no) information from the MND team about respiratory failure/NIV. It is important to recognise that this is what was perceived to be happening by respiratory service-HCPs and therefore not necessarily a true reflection of information provision by the MND team.

Early diagnosis of respiratory dysfunction may prevent the need for urgent/late initiation of NIV, which is associated with reduced compliance and survival [15, 20]. Despite NICE guidelines recommending patients are seen within 1 week of an urgent referral to respiratory/ventilation services [6], nearly two-thirds of respiratory service-HCPs reported that patients usually wait >1 week and some reported waiting times of up to 3 months for routine referrals. Services need to be staffed and flexible to respond to the need for rapid NIV initiation and avoid delays, *e.g.*, considering using outpatient initiation, which has been associated with more rapid initiation and reduced early mortality in patients awaiting an inpatient bed [21].

As outlined in published guidance, regular assessment of respiratory function beginning at, or soon after, diagnosis and using robust measures that are interpreted in combination with symptoms will identify respiratory dysfunction earlier [6–9]. All but one UK service used a combination of respiratory tests to direct clinical decision-making. Despite FVC poorly correlating with respiratory symptoms [22], it is still the most commonly used test. The mean respiratory function thresholds for considering NIV were higher than that recommended by the guidelines reflecting the growing recognition that early initiation of NIV may improve outcomes. However, many staff were using very low thresholds to trigger NIV initiation, which may allow insufficient time for preparation and optimisation of NIV [6]. The more predictive sniff nasal inspiratory pressure was only used by 53% of services [23], although 72% were using blood gases, which are more sensitive [24]. In a recent Italian study, after plwMND were initiated on NIV their bicarbonate levels were a predictor of their adherence and tolerance to NIV as well as death [25]. When bicarbonate levels were above 29 $mmol \cdot L^{-1}$, patients' survival was significantly shortened [25]. We should be mindful that additional/more complex tests may be helpful in difficult cases but may delay diagnosis/initiation of NIV.

Following initiation, evidence suggests that many patients do not reach adequate usage or effective ventilation [3], which is associated with reduced survival [26]. Contrary to guidance [6–9], our findings indicate that monitoring was often infrequent and many relied on subjective measures alone. Telemonitoring provides a solution to receive feedback in real time, but this requires staffing and expertise [27, 28]. Sufficient equipment is needed to optimise patients; however, only 20 services were able to provide a core level of equipment, and out-of-hours support was often limited.

We found that multiple specialists need to be involved to ensure that each component of the pathway is effective in order to optimally deliver NIV [6–10]. This may explain why patients who attend a specialist multidisciplinary centre have improved survival compared to patients attending a non-specialist centre [29]. This coordinated assessment and decision-making process may reduce decision-making delays and facilitate sharing of good practice.

Strengths and limitations

Our findings reflect the current practice of most services delivering NIV in the UK, although there was some variation in responses given by HCPs working in the same service. Therefore, analysis at the service level was based on responses where there was no variation or used the majority response. The study was also self-reported, and at times HCPs reported what they thought services did, which may not necessarily

reflect reality. Similarly, the respondents had a wide range of experience and backgrounds reflecting the usual staff make-up within these services, which may explain some of the variation in responses.

To our knowledge there has been no comparable nationwide service evaluation in the UK or other countries. Therefore, it is important to acknowledge that service variation may be even greater in countries without a publicly funded health service and in low-socioeconomic countries due to the complexity of delivering NIV and the cost/access to equipment.

Our surveys were carried out prior to the COVID-19 pandemic. We recognise that services will have changed due to the pandemic. Factors such as redeployment of staff and equipment, disruption in the multidisciplinary team and difficulties seeing patients face-to-face have impacted upon services [30]. Some respiratory function tests have been identified as aerosol-generating procedures and therefore difficult to conduct. PlwMND have faced longer waiting times for testing/appointments and the provision of treatments have been disrupted [30–32]. The pandemic may pose opportunities to improve services through more experience of remote monitoring, multidisciplinary working becoming more accessible, and more staff having been exposed to using NIV and, therefore, gaining expertise.

Our study focused on respiratory assessment and delivery of NIV, though from our earlier work, we recognise that a holistic approach to respiratory care should include optimisation of cough, secretions and psychosocial matters as well, as they can influence NIV success [4, 5]. Our educational website (www.niv4mnd.co.uk) contains further information on ancillary respiratory care, but there is even less evidence in these areas to guide practice and we recommend that this is a key priority to explore in future research using a similar approach to that adopted in this study.

Conclusion

There is considerable variation in the quality of the NIV service available to patients with MND in the UK. Key issues include delays in the pathway, lack of equipment and variation in staff expertise and behaviour. Good practice appears achievable but is not universally available for every patient. There needs to be increased awareness of the areas of the need for improvement in each service at every stage of the respiratory care pathway. Staff training, improved funding and service reconfiguration may be needed to deliver this.

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References

1 Rafiq MK, Proctor AR, McDermott CJ, *et al.* Respiratory management of motor neurone disease: a review of current practice and new developments. *Pract Neurol* 2012; 12: 166–176.
2 Bourke SC, Tomlinson M, Williams TL, *et al.* Effects of non-invasive ventilation on survival and quality of life in patients with amyotrophic lateral sclerosis: a randomised controlled trial. *Lancet Neurol* 2006; 5: 140–147.
3 McDermott CJ, Bradburn MJ, Maguire C, *et al.* DiPALS: diaphragm pacing in patients with amyotrophic lateral sclerosis – a randomised controlled trial. *Health Technol Assess* 2016; 20: 1–186.
4 Baxter SK, Johnson M, Clowes M, *et al.* Optimizing the noninvasive ventilation pathway for patients with amyotrophic lateral sclerosis/motor neuron disease: a systematic review. *Amyotroph Lateral Scler Frontotemporal Degener* 2019; 20: 461–472.
5 O'Brien D, Stavroulakis T, Baxter S, *et al.* The optimisation of noninvasive ventilation in amyotrophic lateral sclerosis: a systematic review. *Eur Respir J* 2019; 54: 1900261.
6 National Institute for Health and Care Excellence. Motor neurone disease: assessment and management (NICE guideline 42). July 2019. www.nice.org.uk/guidance/ng42 Date last updated: 23 July 2019. Date last accessed: 12 November 2022.

7 Motor Neurone Disease Association. Managing respiratory symptoms in motor neurone disease. September 2017. https://www.mndassociation.org/app/uploads/information-sheet-p6-evaluation-and-management-of-respiratory-symptoms-in-mnd.pdf
8 EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis. EFNS guidelines on the clinical management of amyotrophic lateral sclerosis (MALS): revised report of an EFNS task force. *Eur J Neurol* 2012; 19: 360–375.
9 Miller RG, Jackson CE, Kasarskis EJ, *et al.* Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: drug, nutritional, and respiratory therapies (an evidence-based review): report of the Quality Standards Subcommittee of the American Academy of Neurology. *Neurology* 2009; 73: 1218–1226.
10 Davidson AC, Banham S, Elliott M, *et al.* BTS/ICS guideline for the ventilatory management of acute hypercapnic respiratory failure in adults. *Thorax* 2016; 71: ii1–ii35.
11 Fairbairn S. Wales acute non-invasive ventilation (NIV) guidelines. NHS Wales. https://wales.pallcare.info/files/docs/MND%20pathways%20and%20documents/Saliva%20management%20pathway%20v1.pdf Date last updated: 1 May 2019. Date last accessed: 23 January 2023.
12 Faull C, Oliver D. Withdrawal of ventilation at the request of a patient with motor neurone disease: guidance for professionals. *BMJ Support Palliat Care* 2016; 6: 144–146.
13 Schoonenboom J, Johnson RB. How to construct a mixed methods research design. *Kolner Z Soz Sozpsychol* 2017; 69: Suppl 2, 101–131.
14 Andersen PM, Borasio GD, Dengler R, *et al.* Good practice in the management of amyotrophic lateral sclerosis: clinical guidelines. An evidence-based review with good practice points. EALSC Working Group. *Amyotroph Lateral Scler* 2007; 84: 195–213.
15 Baxter SK, Baird WO, Thompson S, *et al.* The initiation of non-invasive ventilation for patients with motor neuron disease: patient and carer perceptions of obstacles and outcomes. *Amyotroph Lateral Scler Frontotemporal Degener* 2013; 14: 105–110.
16 Martin NH, Lawrence V, Murray J, *et al.* Decision making about gastrostomy and noninvasive ventilation in amyotrophic lateral sclerosis. *Qual Health Res* 2016; 26: 1366–1381.
17 Lechtzin N, Scott Y, Busse AM, *et al.* Early use of non-invasive ventilation prolongs survival in subjects with ALS. *Amyotroph Lateral Scler Frontotemporal Degener* 2007; 8: 185–188.
18 Georges M, Golmard JL, Llontop C, *et al.* Initiation of non-invasive ventilation in amyotrophic lateral sclerosis and clinical practice guidelines: single-centre, retrospective, descriptive study in a national reference centre. *Amyotroph Lateral Scler Frontotemporal Degener* 2017; 18: 46–52.
19 Chio A, Silani V, Italian ALS Study Group. Amyotrophic lateral sclerosis care in Italy: a nationwide study in neurological centers. *J Neurol Sci* 2001; 191: 145–150.
20 Pinto AC, de Carvalho M, Evangelista T, *et al.* Nocturnal pulse oximetry: a new approach to establish the appropriate time for non-invasive ventilation in ALS patients. *Amyotroph Lateral Scler Frontotemporal Degener* 2003; 4: 31–35.
21 Sheers N, Berlowitz DJ, Rautela L, *et al.* Improved survival with an ambulatory model of non-invasive ventilation implementation in motor neuron disease. *Amyotroph Lateral Scler Frontotemproral Degener* 2014; 15: 180–184.
22 Jackson CE, Rosenfeld J, Moore DH, *et al.* A preliminary evaluation of a prospective study of pulmonary function studies and symptoms of hypoventilation in ALS/MND patients. *J Neurol Sci* 2001; 191: 75–78.
23 Polkey MI, Lyall RA, Yang K, *et al.* Respiratory muscle strength as a predictive biomarker for survival in amyotrophic lateral sclerosis. *Am J Respir Crit Care Med* 2017; 195: 86–95.
24 Manera U, Torrieri MC, Moglia C, *et al.* The role of arterial blood gas analysis (ABG) in amyotrophic lateral sclerosis respiratory monitoring. *J Neurol Neurosurg Psychiatry* 2020; 91: 999–1000.
25 Manera U, Torrieri MC, Moglia C, *et al.* Arterial blood gas analysis: base excess and carbonate are predictive of noninvasive ventilation adaptation and survival in amyotrophic lateral sclerosis. *Amyotroph Lateral Scler Frontotemporal Degener* 2021; 22: 33–39.
26 Kleopa KA, Sherman M, Neal B, *et al.* BiPAP improves survival and rate of pulmonary function decline in patients with ALS. *J Neurol Sci* 1999; 164: 82–88.
27 Hobson EV, Baird WO, Bradburn M, *et al.* Using telehealth in motor neuron disease to increase access to specialist multidisciplinary care: a UK-based pilot and feasibility study. *BMJ Open* 2019; 9: e028525.
28 Hobson E, Baird W, Bradburn M, *et al.* Process evaluation and exploration of telehealth in motor neuron disease in a UK specialist centre. *BMJ Open* 2019; 9: e028526.
29 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.
30 Musson LS, Collins A, Opie-Martin O, *et al.* Impact of the Covid-19 pandemic on amyotrophic lateral sclerosis care in the UK. *Amyotroph Lateral Scler Frontotemporal Degener* 2023; 24: 91–99.
31 Glasmacher SA, Larraz J, Mehta AR, *et al.* The immediate impact of the COVID-19 pandemic on motor neuron disease services and mortality in Scotland. *J Neurol* 2021; 268: 2038–2040.
32 Andrews JA, Berry JD, Baloh RH, *et al.* Amyotrophic lateral sclerosis care and research in the United States during the COVID-19 pandemic: challenges and opportunities. *Muscle Nerve* 2020; 62: 182–186.