Results Mean age was 50.6 (21.3). 44% male. 53% used NIV at night. All were naïve to NIV during exercise. NIV increased cycle time by 146 s (105%). All stated they would be prepared to repeat this type of exercise (Abstract P269 table 1).

Conclusion NIV is well tolerated, feasible and significantly increases exercise capacity in patients hospitalised with an acute exacerbation of respiratory disease.

P270 MOTOR NEURONE DISEASE (MND); A SURVEY OF DEATHS IN THE ERA OF NON-INVASIVE VENTILATION

doi:10.1136/thoraxjnl-2011-201054c.270

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Background The care of patients with MND has changed radically with greater uptake of non-invasive ventilation (NIV) and tube feeding (TF). Community colleagues in our region have expressed anxiety about how such supported patients might die and in particular whether the process of dying might be prolonged. We sought to investigate these concerns.

Methods A structured questionnaire was distributed to involved clinicians, community practitioners and MNDA care co-ordinators. They completed questionnaires for patients dying under their care, where necessary/appropriate with the input of family members. The data are compared with an historical, retrospective series (n=50) reported by the Wisdom Hospice (WH).[1]

Results From October 2010 to June 2011, 51 deaths were recorded (mean age 67 years, 29 men). Thirty deaths were in an acute hospital, 15 at home, six in a hospice or community hospital. 16 patients had advance care plans (ACP) of whom 10 died in their "preferred place". Assisted ventilation had been prescribed to 30 and tube feeding to 37 patients. NIV and TF were discontinued before death in 13 and five patients respectively. For 22 patients the process of dying was sudden or <24 h in duration. NIV was not associated with a prolonged process of dying. The commonest certified cause of death was an unqualified "MND", with pneumonia reported in seven cases. No patient had a post mortem examination. The WH cohort had a similar proportion of men and the mean age at death was 66 years. None of the patients had assisted ventilation and only seven had tube feeding. The process of dying was 24 h or less for 24 patients. The commonest recorded cause of death was respiratory failure (22 individuals).

Conclusion These preliminary results show that there is no trend to prolonged deaths in patients with MND using NIV and TF. Several patients have elected to discontinue NIV. ACP's in our region remain patchy and require further attention. The quality of death certification is poor with little detail on the mechanism of death.

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P271 IMPACT OF STRUCTURED REFERRAL AND FOLLOW-UP PATHWAYS ON ACCESS TO VENTILATORY SUPPORT FOR PEOPLE WITH MOTOR NEURONE DISEASE (MND)

doi:10.1136/thoraxjnl-2011-201054c.271

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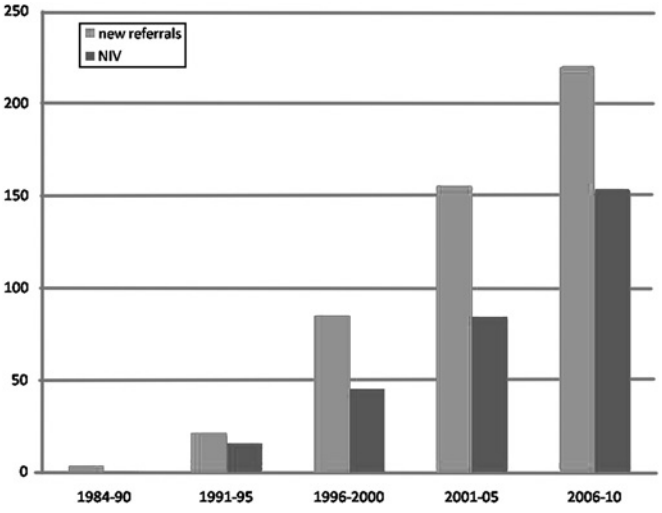
Introduction Attitudes to surveillance for and treatment of ventilatory failure among people with MND have changed over several years. In the UK this culminated in the publication in July 2010 of National Institute of Clinical Excellence (NICE) guidance designed to increase access to non-invasive ventilation (NIV). The services offered in our respiratory unit have evolved on the basis of published evidence in advance of formal guidelines. We examined the impact of compliance with practice parameter recommendations in NICE to our referral numbers and uptake of NIV in East Anglia.

Methods A retrospective review of number of referrals and new NIV starters from 1984 to 2010 in a regional respiratory support unit. Between 2001 and 2005 the MND Association helped to establish a care centre in Cambridge and closer working was established with the respiratory service. From 2006 the default position was to offer all patients newly diagnosed with MND a respiratory assessment and structured 3 monthly follow-up appointments in a fashion subsequently detailed in the NICE guidance.

Results The numbers of new referrals and people starting NIV are shown in Abstract P271 figure 1. Between 1984 and 2000 there was slow growth but the mean annual values were just seven referrals and four new NIV starters (57%). With closer working between neurologists in the care centre and the respiratory unit between 2001 and 2005 mean referral numbers increased to 31 with 17 new NIV starters (55%) per year. With default referral and 3 monthly reviews the number referred grew to 44 with around 31 NIV starters per year (70%).



Abstract P271 Figure 1

Conclusion With an estimated population of 2.5 million in East Anglia, and an annual incidence of 2.8 per 100 000 we estimate 70 new cases of MND per year. Around 60% of patients are therefore being referred and 70% of these are starting NIV. Implementing the NICE guidance could have a similar impact across the UK, a great improvement on the position in 2000 when only around 3% of patients were being offered NIV.[1]

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P272 A 7 YEAR RETROSPECTIVE EVALUATION OF INITIATION OF LONG TERM NON-INVASIVE VENTILATORY SUPPORT FOR MOTOR NEURONE DISEASE

doi:10.1136/thoraxjnl-2011-201054c.272

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In 2010 NICE (1) published it's guideline for the use of non-invasive ventilation in the management of motor neurone disease (MND). We have offered long-term non-invasive ventilation (NIV) in this

patient group for7 years. We have evaluated concordance data for all patients set-up during this time.

Methodology Data were obtained by retrospective case-note review of all patients set-up on NIV for MND between April 2004 and March 2011inclusive.

Results 42 patients were set-up over 7 years. Mean length of time from diagnosis to ventilation was 13.9 months, with three patients being set-up on diagnosis and a further three before diagnosis. 69% (n=29) were set-up at home, the remainder in hospital. 71% (n=30) of those set up were eventually concordant, 19% (n=8) did not tolerate NIV, while 10% (n=4) died during initiation. Those set-up in hospital had a tendency to be more concordant with ventilation than those set-up at home (76% vs 69%). This group also became concordant more rapidly (4.4 days vs 14.2 days). Those with both symptomatic and physiological indications for ventilation appeared to tolerate it extremely well with 84% (n=24) becoming concordant. There was a failure rate of 75% of those with physiological indications only. Of those with symptoms only and no apparent abnormal physiological markers 80% (n=4) complied with treatment. Symptomatically daytime somnolence was the most commonly reported symptom, with 81% (n=34) of patients being sleepy by day. Nocturia was the least commonly reported at 11%. In terms of mortality, mean survival from initiation to death was 10.2 months (range 0.67–84). In total three patients progressed from NIV to tracheostomy ventilation, one of these survived a further 5 years.

Conclusion Those with MND have a tendency to have better concordance with NIV if started in hospital rather than at home; concordance also appears to be more rapid in this group. As expected, given the nature of this treatment those without symptoms tend not to tolerate NIV.

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P273 OUTCOME AFTER PROLONGED INVASIVE MECHANICAL VENTILATION IN MYOTONIC DYSTROPHY

doi:10.1136/thoraxjnl-2011-201054c.273

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Introduction Patients with myotonic dystrophy (MD) may present acutely with respiratory failure due to muscle weakness and aspiration pneumonia. We are not aware of published data regarding the outcome of intubation and ventilation in these circumstances.

Method We performed a retrospective case notes review of patients referred to a specialist respiratory support centre with a diagnosis of MD. In this abstract we present the outcomes of patients who had received invasive mechanical ventilation and were transferred as in-patients to our unit.

Results Between 1996 and 2010, 22 patients (seven men), mean age 39 years, who had all been intubated and ventilated for a minimum of 2 weeks, were referred. At transfer, 16 patients were tracheostomy ventilated, two were on non-invasive ventilation (NIV) and five were self-ventilating but had on-going CO_2 retention. Where specified, admission to ICU was precipitated by pneumonia (16), sudden collapse (2) and cardiac failure (2). The mean duration of invasive ventilatory support was 67 (range 17–196) days. One patient died in our unit. Of those discharged, 11 were on NIV, five still required tracheostomy ventilation and six were ventilator independent. Five patients required a mini-trach tube for access to secretions. The mean length of stay in the weaning unit was 35 (range 2–95) days. Mean survival post discharge was 44 (range 1–102) months. In the same period 53 patients with MD who had not been intubated and ventilated were initiated on NIV for ventilatory failure in our unit. Mean survival in this group was 56 (range 1–144) months.

Conclusion Patients with MD may require prolonged invasive ventilation when presenting with acute ventilatory decompensation. In this series the majority weaned from invasive mechanical ventilation and treatment including NIV was associated with survival on average of over 3.5 years. The prognosis may not be as good as for those patients with MD starting NIV who have not required prior intubation.

P274 SEASONAL VARIATION IN INITIATION AND DISCONTINUATION OF DOMICILIARY NON-INVASIVE VENTILATION: A 12-MONTH COHORT STUDY

doi:10.1136/thoraxjnl-2011-201054c.274

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Introduction Domiciliary non-invasive ventilation (NIV) is an established treatment for patients with chronic respiratory failure. Although the proportion of patients that discontinue NIV is reported from clinical trial data, observational cohort studies often lack these details. Furthermore, adherence to treatment and patient retention is enhanced in clinical trials as a consequence of well-defined patient selection and greater clinical and non-clinical support for trial patients.

Aim To investigate initiation, discontinuation and rationale for discontinuation in patients initiated on domiciliary NIV in a regional home mechanical ventilation centre over a 12-month period.

Methods All data were collected prospectively from a discharge summary database form 1 January to 31 December 2010. Monthly initiation and discontinuation trends and rationale for discontinuation were analysed across differing diagnostic groups.

Results 200 patients were initiated (123) and discontinued (77). Chronic obstructive pulmonary disease (COPD), neuromuscular and chest wall disease (NMD and CWD) and obesity related respiratory failure (ORRF) were the most frequent diagnoses for initiation (26.0%, 23.6%, 50.4%, respectively) and discontinuation (13.8%, 22.8%, 41.6%, respectively). Overall initiation rates were constant throughout the year with a fall in the number of COPD patients during the Summer. Death (52.6%) and poor adherence to the ventilator prescription (19.2%) were the commonest reasons for discontinuation across all groups. As expected, death was the commonest indication for discontinuation in the COPD and NMD and CWD group and poor adherence in the ORRF group (Abstract P274 table 1).

Abstract P274 Table 1 Initiation and discontinuation of domiciliary NIV

	Initiation n (%)				Termination n (%)			
	COPD	NMD and CWD	ORRF	Total	COPD	NMD and CWD	ORRF	Total
Spring	9 (28)	9 (31)	17 (27)	35 (28)	7 (41)	9 (32)	11 (34)	27 (35)
Summer	4 (13)	6 (21)	19 (31)	29 (24)	3 (18)	5 (18)	6 (19)	14 (18)
Autumn	10 (31)	9 (31)	13 (21)	32 (26)	2 (12)	5 (18)	8 (25)	15 (19)
Winter	9 (28)	5 (17)	13 (21)	27 (22)	5 (29)	9 (32)	7 (22)	21 (27)
Total	32 (100)	29 (100)	62 (100)	123 (100)	17 (100)	28 (100)	32 (100)	77 (100)

Discussion Although there is seasonal variation in the initiation of NIV in the COPD patients, this is not apparent in the NWD, CWD and ORRF patients. However, there is a seasonal variation in stopping NIV, which may relate to excess mortality in the Winter