

patient group for 7 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 2011 inclusive.

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.

REFERENCE

1. NICE. Motor Neurone Disease: The Use of Non-Invasive Ventilation in the Management of Motor Neurone Disease. London: NICE, 2010.

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 inpatients 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 CO₂ 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 venti-

lated 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 decompression. 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.

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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 from 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		Total	COPD	NMD and CWD		Total
		ORRF	Total			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